TRANSITIONAL CARE
IN YOUNG PEOPLE WITH CONGENITAL HEART DISEASE
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TRANSITIONAL CARE IN YOUNG PEOPLE WITH CONGENITAL HEART DISEASE

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you have taught me to be courageous

to believe in my dreams

even after a million tomorrows

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LIST OF ABBREVIATIONS

ACC	American College of Cardiology
ACHD	Adult Congenital Heart Disease
ANOVA	Analysis of Variance
APN	Advanced Practice Nursing
ASD	Atrial Septal Defect
AUDIT	Alcohol Use Disorders Identification Test
AVSD	Atrio-Ventricular Septal Defect
CAH	Congenital Adrenal Hyperplasia
CCC	Complex Chronic Condition
CCU	Cardiac Care Unit
CHD	Congenital Heart Disease
CIs	Confidence Intervals
CONCOR	CONgenital COR Vitia
DI	Disagreement Index
DILV	Double Inlet Left Ventricle
DORV	Double Outlet Right Ventricle
ECG	Electrocardiogram
EPS	Education and Psychosocial Support
ESC	European Society of Cardiology
GLM	General Linear Model

GP	General Practitioner
GRI	Guyatt's Responsiveness Index
HBS-CHD	Health Behavior Scale – Congenital Heart Disease
HIV	Human Immunodeficiency Virus
ICU	Intensive Care Unit
I-CVI	Item Content Validity Index
i-DETACH	Information Technology Devices and Education program for Transitioning of Adolescents with Congenital Heart disease
IE	Infective Endocarditis
INTERCHANGE	INTERnational study on the Continuation of Heart health checks in young adults with congenital heart disease
IPR	Interpercentile Range
IPRAS	Interpercentile Range Adjusted for Symmetry
IRB	Institutional Review Board
ISACHD	International Society for Adult Congenital Heart Disease
JIA	Juvenile Idiopathic Arthritis
LKQ-CHD	Leuven Knowledge Questionnaire – Congenital Heart Disease
MINORS	Methodological Index for Non-Randomnized Studies
ORs	Odds Ratios
PhD	Philosophiae Doctor
RCT	Randomnized Controlled Trial
RIDIT	Relative to an Identified Distribution
RR	Response Rate

SCD	Sickle Cell Disease
S-CVI/Ave	Scale Content Validity Index/Average
SD	Standard Deviation
SWITCH^2	Self-management and Well-being Improvements by Transitioning adolescents with Chronic disorders in Hospitals and at Home
TGA	Transposition of the Great Arteries
UK	United Kingdom
USA	United States of America
VSD	Ventricular Septal Defect
Quan-Qual	Quantitative-qualitative
Q1	First quartile
Q3	Third quartile
XTC	ecstasy
YRBS	Youth Risk Behaviors Survey
ZIP	Zone Improvement Plan

1 GENERAL INTRODUCTION

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Congenital heart disease (CHD) is defined as anatomical defects of the heart or intrathoracic great vessels that are actually or potentially of functional significance (1). It comprises a wide spectrum of simple, moderate, and complex heart lesions. This condition has an estimated birth incidence of 9 per 1,000 newborns and continues to affect most patients throughout their lives. Thanks to advances in diagnostic capacities, therapies, and critical care, the life expectancy of these children increased substantially. Nowadays, about 90% of children born with a heart lesion reach adult age (2). Despite these improved prospects and irrespective of the initial treatment, most patients with CHD cannot be considered as cured. These patients remain at risk for developing complications such as heart failure, endocarditis, and arrhythmias throughout their entire life spectrum. In order to prevent these complications, life-long follow-up and care is required. Indeed, as CHD became a life-cycle disease, patients move from pediatric cardiology, over adolescent clinics, to adult-focused settings and even geriatric care (3). At each life stage an age-appropriate care program is required to address the specific medical and non-medical needs of this population.

During childhood, patients should receive care at specialized pediatric cardiology programs. When reaching adulthood, care should be provided at different levels according to the heart defect and related care needs. According to expert-based international consensus statements, care can be provided at three different levels; more specifically in adult congenital heart disease (ACHD) programs located in tertiary care centers; non-specialist care settings (e.g., general cardiology care); or in shared care programs (4;5). International guidelines state that during the developmental transition of young people with CHD towards adulthood, provision of continuous, uninterrupted care is mandatory. Therefore, in most cases, a timely transfer from pediatric to adult-centered care is essential. However, in some healthcare settings specific care programmes are provided to patients with CHD 'from birth until death'. In these latter programs, an actual transfer of care from pediatric to adult-centered care is not mandatory, since patients are provided continuous care at each age or life stage. (5-7). Over the past decade, the issues of transfer and transition have received increasing attention (5-10).

Definitions

Although numerous definitions of transfer and transition can be found in the literature, to date, there is unfortunately no standard definition of these concepts available. Neither is there a clear distinction with or between other related constructs, such as transition process, transition planning, transition readiness, etc. (11). Both in literature and clinical practice, these concepts are frequently, although incorrectly, used interchangeably.

Transfer has been defined as "*an event or series of events through which adolescents and young adults with chronic physical and medical conditions move their care from a pediatric to an adult healthcare environment*" (12). The goal of transfer of care is to maximize patient's functioning through the provision of high-quality, developmentally appropriate, continuous healthcare services (13). In late adolescence, a pediatric setting may be less suitable to provide (age-)appropriate care. Therefore, it is recommended to transfer the care for patients from a pediatric to an adult-focused setting when patients reach adult age where one is available (14). More specifically, patients with CHD ought to be transferred from pediatric cardiology to an Adult Congenital Heart Disease (ACHD) program, understanding that this may not be feasible in areas not having such an ACHD center.

Transitions, in general, are passages from one life phase, physical condition, or social role to another (15). Four types of transition have been previously described (16). First, 'health/illness transitions' refer to changes in health status of patients and range from adapting to a chronic illness, returning home from hospitalization, or recovering from surgery. Second, 'developmental transitions' occur with standard changes in the developmental stages of life such as adolescence, parenthood or aging. Third, 'situational transitions' pertain to environmental, contextual, and social changes, such as changing educational or professional roles or altered family situations. Fourth, 'organizational transitions' reflect changes in leadership, policies or organizational structures, affecting both personnel and clients of an organization (16).

With respect to the provision of adolescent health care, developmental transitions are the most relevant type of transition. From this perspective, transition can be viewed as the evolution of an individual with a chronic condition from a dependent child to an independent adult. It corresponds with "*the process by which adolescents and young adults with chronic childhood illnesses are prepared to take charge of their lives and their health in adulthood"* (12). Transition as a healthcare intervention is frequently defined as "*a purposeful, multi-faceted, planned process that addresses the medical, psychosocial, and educational/vocational needs of adolescents and young adults with chronic physical and medical conditions"* (17). All too often, it is added that this process is undertaken "*as patients move from child-centered to adult-oriented healthcare systems"*. However, this additional statement may confuse the distinction from the concept of transfer. Furthermore, it is frequently argued that the transition process is needed to prepare patients for the transfer from pediatric to adult care (10;12). However, transition should not end at the time of transfer, because the individual continues to evolve towards adulthood (12). Indeed, transitional care remains important to further coach and guide individuals with CHD towards adulthood, even beyond the

event of transfer to adult care. Among young adults who remain with a pediatric cardiologist into adulthood (i.e., do not undergo transfer of care), transition remains an important and necessary process. Indeed, transition is normal and something that every adolescent or young adult with CHD needs to go through.

Transfer

Transfer of care from a pediatric to an ACHD program allows patients with CHD to be cared for in an environment suiting their medical and psychosocial needs. For example, adult survivors of CHD may develop comorbidities with which pediatricians are not familiar. Healthcare transfer overlaps, however, also with other significant life events, such as independent living, employment, relationships, and family planning, which adult providers are better suited to address. From a practical perspective, pediatric hospitals in many areas have an upper age limit (typically 16-18 years) for inpatient care. For these reasons, the involvement of adult specialists, where available, is mandatory for the overall well-being of young adults with CHD, as is true for young adults living with other types of chronic health conditions.

Not only is a transfer of care important from the perspective of providing high-quality care, it is also important to many young people. Although data are mixed, with many young adults being reluctant to leave the comfort of the familiar pediatric setting (18), others indicate a lack of enthusiasm for continuing to share a waiting room with infants and toddlers (19), feeling that the experience of attending the pediatric cardiology clinic is something that they have “outgrown”.

The organization of transfer of care

Numerous expert-based recommendations emphasize the need to develop structured and flexible plans for the transfer of care from pediatric cardiology to an adult-focused care setting (8-10;20). These recommendations indicate that transfer of care, or the actual handing-off of the responsibility of care to the patient and a team of ACHD providers, should be part of a comprehensive transition program (8;10).

The transfer of care should be made towards the recommended level of ACHD care based on an established algorithm for the initial evaluation and ongoing follow-up of adults with CHD (20). It is

recommended that every patient, irrespective of the level of disease complexity, should be seen by an ACHD specialist at least once after the transfer from pediatric cardiology. During this initial post-transfer evaluation, a thorough clinical assessment of the patient's overall status is performed in order to perform a risk stratification and decide on the advised setting, level, and frequency of future follow-up visits (20). Hence, this initial visit to a regional ACHD center provides a good opportunity for the care team to review patient's understanding about the heart lesion(s), anticipated prognosis including the possible need for (redo-)interventions in the future, the risk of developing complications, and strategies to access adult healthcare, especially in urgent situations (9;20).

Based on international expert opinion, the recommended levels of follow-up are:

Level 1 – Specialist care: Care exclusively performed by specialized CHD cardiologists in regional tertiary ACHD clinics (9;20) or at satellite centers. Satellite centers are regional hospitals in which a CHD cardiologist performs outpatient visits (21). This level of care is indispensable for patients with complex cardiac conditions (9).

Level 2 – Shared care: Shared care is defined as care performed by a general cardiologist in collaboration with and/or by sending reports to specialist centers (9;21). Patients with mild to moderate complexity lesions can be seen in shared care facilities (9).

Level 3 – Non-specialist care: Non-specialist cardiac care refers to follow-up provided by a general adult cardiologist but without a formal collaboration with an ACHD program; or care provided by a general practitioner or family physician (9;20;21). This level of care is appropriate for patients who are at low risk for developing comorbidities or long-term complications (9;20). These are typically mild heart defects. Access to specialized ACHD care must, however, be possible at any time if needed.

Table 1.1: Overview of primary heart defects with corresponding level of anatomical complexity

Congenital Heart Lesions: Primary diagnosis	Level of anatomical complexity based on Task Force 1
Hypoplastic left heart syndrome	complex
Univentricular physiology	complex
Tricuspid atresia	complex
Tetralogy of Fallot	moderate
Pulmonary atresia with VSD	complex
Pulmonary atresia without VSD	complex
Double Outlet Right Ventricle	complex
Double Inlet Left Ventricle	complex
Truncus arteriosus	complex
Transposition of the Great Arteries (TGA)	complex
Congenitally Corrected-TGA	complex
Coarctation of the aorta	moderate
Atrioventricular Septal Defect	moderate
Atrial Septal Defect type 1	moderate
Ebstein malformation	moderate
Pulmonary Valve abnormality	moderate to complex
Aortic valve abnormality	simple if isolated, moderate if associated
Aortic abnormality	moderate
Left ventricle outflow tract obstruction	moderate
Atrial Septal Defect type 2	simple if repaired without residua, moderate if associated
Ventricular Septal Defect (VSD)	simple if repaired without residua, moderate if associated
Mitral valve abnormality	simple if isolated, moderate if associated
Pulmonary vein abnormality	moderate

Despite the well-established rationale for transferring patients to adult care, only three-quarters of pediatric cardiology programs in Europe and North America appeared to transfer their patients to adult-focused care, and only one in two transfer patients to formalized ACHD programs (22). These data were published in 2009 and the situation may have improved in the interim. Nonetheless, it is clear that ACHD guidelines and review articles published before this study had not led all pediatric cardiology programs to adopt transfer of care (12;23;24).

A prerequisite of transferring adolescents to ACHD programs is that they have not yet left the healthcare system (i.e., become lost to follow-up) before reaching the age of transfer. Likewise, the process of transfer needs to be seamless from the patient's perspective in order to not discourage young adults from attending their first ACHD appointment. Unfortunately, loss to follow-up is a common problem in CHD and occurs both within the pediatric age range, as during and after the transfer process. Reid and colleagues were the first to report on the scope of the problem of loss to follow-up (25). Among 360 young adults (19-21 year) with moderate or complex CHD, only 47% had attended an ACHD clinic after the transfer of care (25). This occurred in the context of a universal healthcare system free of financial barriers to care. Mackie and colleagues demonstrated how increasing age is associated with a steady decline in attendance at pediatric cardiology clinics, with >20% of youth with complex lesions lost to follow-up before reaching adult age (26). Loss to follow-up was even more prevalent among subjects having less severe lesions but requiring periodic cardiology reassessments (26). As a result, less than half of the adults in Canada who require ACHD care are actually followed in such centers (27). Yeung and colleagues reported on 158 adults with moderate or complex CHD referred to an ACHD program in Colorado (USA), of whom 99 (63%) had a lapse in care of more than two years since being seen in a pediatric cardiology clinic (28). One third had been told there was no need for follow-up (28), despite guidelines to the contrary (29).

To date, only a limited set of risk factors for loss to follow-up has been identified, including male sex (26), lower socio-economic status, absence of chart documentation regarding the need for follow-up (25;30), and fewer surgical interventions (25). Interview data has identified additional factors contributing to loss to follow-up, including a lack of awareness of the importance of follow-up and lack of an organized approach within some families to remembering appointments (30). The presence of comorbid conditions, avoidance of risk-taking behaviors, and a history of attending pediatric cardiology appointments without a parent or sibling were factors found to be associated with a successful transfer (31).

In order to bring about a change in practice, outcome data among young adults who experienced care gaps is needed. Among 52 Danish adults with CHD who sought and obtained cardiology care in response to an article in the lay press about loss to follow-up, 62% had significant residual lesions and over one third had symptoms at presentation in the adult clinic (32). A study performed by Yeung and colleagues reported that care gaps were associated with an increased risk of cardiac symptoms at presentation and with a three-fold greater likelihood of needing a catheter or surgical intervention within six months of being seen at the adult care facility (28). As important as

these experiences are, more outcome data are highly needed with respect to transfer from pediatric to adult care, and in particular to identify which clinical practices best promote successful transfer of care.

Transition

When working with 'adolescents with CHD,' it is likely more natural for healthcare providers to focus on the 'with CHD' qualifier and pay less attention to the 'adolescent' role. From developmental and psychological perspectives, however, adolescents are quite different from children and adults and represent a distinct group that warrants one's attention and respect. Regardless of the presence of a chronic medical condition, a number of changes occur in adolescence, including those related to cognition, emotions, sexuality, and identity (33;34). As individuals progress through adolescence, they move from concrete thinking to more complex abstract thinking and toward social autonomy (34).

Healthcare transition does not occur in isolation and should be considered within the broader context of other developmental transitions common to adolescence and young adulthood, including education, employment, and social relationships. There are distinct developmental tasks that occur during adolescence and others that take place during young adulthood (8). For example, adolescent tasks include setting educational and vocational goals whereas typical tasks during adulthood include obtaining and maintaining employment. With regard to social and family relationships, peers play a prominent role for adolescents, while the selection of life partners and reproductive issues become more prominent in adulthood. These patterns are present for most individuals, irrespective of the presence of CHD. Adolescents and young adults with CHD, however, might benefit from additional support because individuals with pediatric-onset chronic medical conditions are known to achieve fewer developmental milestones (e.g., autonomy, psychosexual development, social development) or achieve these milestones at a later age than healthy peers (35;36).

Furthermore, adolescence is the developmental stage in which people typically challenge authority and take risks (34;37). Risk-taking increases between childhood and adolescence and subsequently decreases between adolescence and adulthood (37). Adolescents take risks partially to establish peer acceptance and autonomy and to experiment with choices (38). Egocentric thinking and the sense of invincibility ('it won't happen to me') are strongest during adolescence (39). Risky health behaviours are certainly not uncommon among adolescents with CHD, with over one-quarter

reporting cigarette smoking, binge drinking, or illicit drug use in a 30 day recall period (40). Furthermore, many young people with CHD have overly optimistic expectations regarding their life expectancy (25). Although risk-taking during adolescence is understandable from a developmental perspective, parents and healthcare providers can support adolescents with CHD to mature and develop effective long-term health behaviours. Positive relationships and having personal goals might also serve as protective factors to lessen involvement in risky behaviours (39).

Despite the well-established importance of developing, implementing, and testing of comprehensive transition programs (10;12;41;42), currently there are minimal empirical data regarding transitional care of young people with CHD. As indicated before, three-quarters of American and European pediatric cardiology programs reported that they transfer patients from pediatric to adult care at an average age of 18 years (22). Of those that transfer patients to adult care providers, less than one-third reported that they provide structured preparation within the form of transition for patients and family (22). Therefore, there is clear discordance between recommendations for a structured approach to transition and what is currently being provided to patients with CHD.

The organization of transition

The general objective of a transition program is to provide adolescents diagnosed with a chronic condition the time, support, and possibility to develop a set of skills, attitudes, and behaviors needed to prepare for adult life. Furthermore, a transition program aims more specifically, to prepare young patients for the transfer of care to an adult-focused program(10;14). A transition program aims to support patients in accomplishing skills related to decision-making, self-advocacy, and self-efficacy (43). Furthermore, transitional care aims to result in uninterrupted, patient-centered, age and developmentally appropriate, comprehensive care. Ultimately, the implementation of a transition program should result in the optimization of the everyday functioning of the adolescent, satisfaction with life, and future productivity in adult life (10;17).

In 2011, recommendations about managing the transition to adulthood of adolescents with CHD were published by the American Heart Association (AHA) (10). This comprehensive document recommends a formal transition process to prepare young people with CHD to become responsible for their health care in order to achieve optimal quantity and quality of life.

Based on these recommendations, transitional care should start at an early age (e.g., 10-12 years) with a pre-transition phase (7). This preparatory phase, generally established in pediatric cardiology, aims to introduce and explain the objectives and key elements of transition and transfer early in childhood. At a young age, it is appropriate to raise the awareness of the patient and parents about the implications of the heart lesion(s), the need for life-long specialized follow-up, and the importance of maintaining a heart-healthy lifestyle. Furthermore, such a pre-transition phase provides parents the opportunity to gradually adapt to the growing independence of their adolescent child (10).

When an adolescent is provided care as part of a transition program, three components are generally included: education and skills development, counseling, and assessment of transition readiness (44;45).

An important component of a transition program is a comprehensive educational curriculum for patients and parents regarding the condition, treatment, preventive measures, and lifestyle matters (10). Several educational topics should be included in this curriculum (5-7;46) such as: the name, description and anatomy of the heart defect; medication regimen; importance of medication adherence; prevention of complications; signs of deterioration needing medical assessment; preventive strategies of endocarditis; need for life-long follow-up; healthy lifestyle choices; heart-healthy sexual behaviors; maternal and fetal risk of pregnancy; and recurrence rates of heart defect(s). Although previous research revealed that significant gaps remain in the knowledge of patients regarding their heart defect (47-56), limited evidence is available supporting the effectiveness of educational interventions in young people with CHD (57-59). Furthermore, it remains currently unknown if the provision of education will result in improvement of the health behaviors of young people with CHD.

Furthermore, a second component of transitional care is supporting patients in developing self-management skills needed to gradually assume responsibility for their health and health care. Self-management skills to be included in the transition curriculum are: scheduling follow-up appointments independently from parents, coordinating prescription renewals, be actively engaged in open communication with healthcare professionals during outpatient visits, being able to discuss topics of concern with providers, and knowing how to obtain and maintain healthcare and health insurance (8-10;12). Finally, issues requiring counseling include education and employment;

disclosing the heart defect to any future employers and colleagues; sexuality, pregnancy and family planning (10).

Finally, at the end of the transition phase, patients' readiness to function as an autonomous adult should be assessed. This includes evaluating whether an adolescent can meet the expectations of society and the adult healthcare system before formally being transferred to an ACHD program (10). It is generally assumed that patients should be capable of planning their own check-ups; be responsible for adhering to the prescribed medication regimen and health behaviors; be able to recognize signs and symptoms of complications; and understand the nature, prognosis, and implications of their condition (10). Assessing the level of self-management and self-efficacy of adolescents with regard to their care is mandatory in order to evaluate the patient's readiness to transfer care (14;43;60-62).

Research objectives

Irrespective the current insights, there remain blind spots in the body of knowledge regarding transfer and transition of adolescents and young adults with CHD. Therefore, this PhD dissertation aimed to address three main objectives (see **Figure 1.1**).

The first objective was to determine the relevance of investigating the concepts of transfer and transition in a population of young people with CHD, and the magnitude of the problem. This objective was addressed in several research questions investigated as part of this PhD:

1. What is the importance of studies investigating transfer and transition on the ACHD nursing research agenda? (Chapter 2)
2. What are the destinations of transfer in adolescents and young adults with CHD leaving pediatric cardiology? (Chapter 3)
3. How are the guidelines for the management of ACHD follow-up care implemented in practice? (Chapter 4)

The second objective was to identify multilevel predictors of care gaps within a broad population of young patients diagnosed with a complex chronic condition from existing literature.

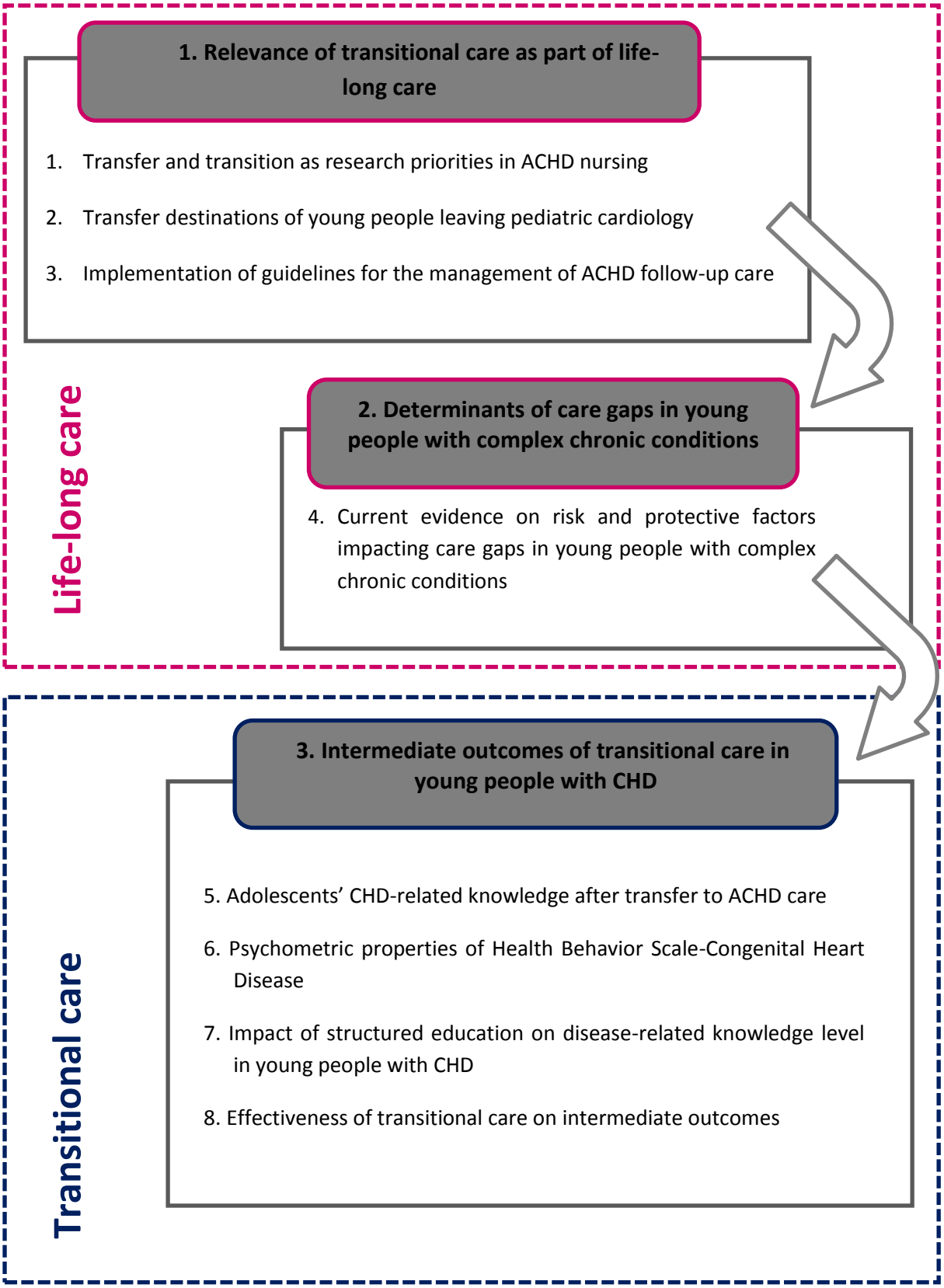
This objective was addressed in a systematic literature review focusing on the following research question:

4. What is the current evidence base on multilevel factors that have an impact on the occurrence of care gaps in adolescents and young adults with complex chronic conditions? (Chapter 5)

Finally the third objective was to assess intermediate outcomes (i.e., disease-related knowledge and health behaviors) of transitional care provision in a sample of adolescents with CHD. A set of subsequent research questions was investigated to address this last aim:

5. What is the adolescents' understanding and knowledge of their heart defect after their transfer to ACHD care? (Chapter 6)
6. What are the psychometric properties of the Health Behavior Scale-Congenital Heart Disease? (Chapter 7)
7. What is the impact of a structured education program on the disease-related knowledge level of young people with CHD? (Chapter 8)
8. What is the effectiveness of transitional care on the disease-related knowledge and health behaviors of young people with CHD? (Chapter 9)

Figure 1.1: Overview of research objectives addressed in this PhD dissertation



References

(1) Mitchell SC, Korones SB, Berendes HW. Congenital heart disease in 56,109 births. Incidence and natural history. Circulation 1971;43(3):323-332.

(2) Moons P, Bovijn L, Budts W, Belmans A, Gewillig M. Temporal Trends in Survival to Adulthood Among Patients Born With Congenital Heart Disease From 1970 to 1992 in Belgium. Circulation 2010;122(22):2264-2272.

(3) Baumgartner H, Budts W, Chessa M, Deanfield JE, Eicken A, Holm J et al. Recommendations for organization of care for adults with congenital heart disease and for training in the subspeciality of 'Grown-up Congenital Heart Disease' in Europe: a position paper of the working Group on Grown-up Congenital Heart Disease of the European Society of Cardiology. Eur Heart J 2014;35(11):686-690.

(4) Baumgartner H, Bonhoeffer P, De Groot NMS, de Haan F, Deanfield J, Galie N et al. ESC Guidelines for the management of grown-up congenital heart disease (new version 2010). Eur Heart J 2010; 31(23):2915-2957

(5) Deanfield J, Thaulow E, Warnes C, Webb GD, Kolbel F, Hoffman A. et al. Management of Grown Up Congenital Heart Disease. The Task Force on the Management of Grown Up Congenital Heart Disease of the European Society of Cardiology. Eur Heart J 2003;24:1035-1084.

(6) Foster E, Graham TP, Driscoll DJ, Reid GJ, Reiss JG, Russell IA et al. Task Force 2: Special health care needs of adults with congenital heart disease. J Am Coll Cardiol 2001;37(5):1176-1183.

(7) Sable C, Foster E, Uzark K, Bjornsen K, Canobbio MM, Connolly HM et al. Best Practices in Managing Transition to Adulthood for Adolescents with Congenital Heart Disease: The Transition Process and Medical and Psychosocial Issues: A Scientific Statement From the American Heart Association. Circulation 2011;123:1454-1485.

(8) Foster E, Graham TP, Driscoll DJ, Reid GJ, Reiss JG, Russell IA et al. Task Force 2: Special health care needs of adults with congenital heart disease. J Am Coll Cardiol 2001;37(5):1176-1183.

(9) Deanfield J, Thaulow E, Warnes C, Webb GD. Kolbel F, Hoffman A et al. Management of Grown Up Congenital Heart Disease. The Task Force on the Management of Grown Up Congenital Heart Disease of the European Society of Cardiology. Eur Heart J 2003;24:1035-1084.

(10) Sable C, Foster E, Uzark K, Bjornsen K, Canobbio MM, Connolly HM et al. Best Practices in Managing Transition to Adulthood for Adolescents with Congenital Heart Disease: The Transition Process and Medical and Psychosocial Issues: A Scientific Statement From the American Heart Association. Circulation 2011;123:1454-1485.

(11) Schwartz LA, Tuchman LK, Hobbie WL et al. A social-ecological model of readiness for transition to adult-oriented care for adolescents and young adults with chronic health conditions. Child Care Health Dev 2011;37(6):883-895.

(12) Knauth A, Verstappen A, Reiss J, Webb GD. Transition and transfer from pediatric to adult care of the young adult with complex congenital heart disease. Cardiol Clin 2006;24(4):619-629.

(13) Moons P, Hilderson D, Van Deyk K. Implementation of transition programs can prevent another lost generation of patients with congenital heart disease. Eur J Cardiovasc Nurs 2008;7(4):259-263.

(14) Rosen DS, Blum RW, Britto M, Sawyer SM, Siegel DM. Transition to adult health care for adolescents and young adults with chronic conditions - Position paper of the Society for Adolescent Medicine. J AdolescHealth 2003;33(4):309-311.

(15) Schumacher KL, Meleis AI. Transitions: a central concept in nursing. Image J Nurs Sch 1994;26:119-127.
(16) Meleis AI, Trangenstein PA. Facilitating Transitions - Redefinition of the Nursing Mission. Nursing Outlook 1994;42(6):255-259.
(17) Blum RW, Garell D, Hodgman CH, Jorissen TW, Okinow NA, Orr DP et al. Transition from child-centered to adult health-care systems for adolescents with chronic conditions. A position paper of the Society for Adolescent Medicine. J Adolesc Health 1993;14(7):570-576.
(18) Moons P, Bovijn L, Budts W, Belmans A, Gewillig M. Temporal trends in survival to adulthood among patients born with congenital heart disease from 1971 to 1992 in Belgium. Circulation 2010;122(22):2264-2272.
(19) Rempel G, Macgill-Evans J, Norris C et al. Ready or not: parenting adolescents with complex congenital heart disease prior to transfer to adult care. Can J Cardiol 2009;25(Suppl SB):245B.
(20) Landzberg MJ, Murphy DJ, Davidson WR, Jarcho JA, Krumholz HM, Mayer JE et al. Task Force 4: Organization of Delivery Systems for Adults with Congenital Heart Disease. J Am Coll Cardiol 2001;37(5):1187-1193.
(21) Goossens E, Stephani I, Hilderson D, Gewillig M, Budts W, Van Deyk K et al. Transfer of Adolescents With Congenital Heart Disease From Pediatric Cardiology to Adult Health Care. An Analysis of Transfer Destinations. J Am Coll Cardiol 2011;57(23):2368-2374.
(22) Hilderson D, Saidi AS, Van Deyk K, Verstappen A, Kovacs AH, Fernandes SM et al. Attitude toward and current practice of transfer and transition of adolescents with congenital heart disease in the United States of America and Europe. Pediatr Cardiol 2009;30(6):786-793.
(23) American Academy of Pediatrics, American Academy of Family Physicians and American College of Physicians-American Society of Internal Medicine. A consensus statement on health care transitions for young adults with special health care needs. Pediatrics 2002;110(6Pt2):1304-1306.
(24) Shaw KL, Southwood TR, McDonagh JE. Development and preliminary validation of the 'Mind the Gap' scale to assess satisfaction with transitional health care among adolescents with juvenile idiopathic arthritis. Child Care Health Dev 2007;33(4):380-388.
(25) Reid GJ, Irvine MJ, McCrindle BW, Sananes R, Ritvo PG, Siu SC et al. Prevalence and correlates of successful transfer from pediatric to adult health care among a cohort of young adults with complex congenital heart defects. Pediatrics 2004;113(3 Pt 1):e197-e205.
(26) Mackie AS, Ionescu-Ittu R, Therrien J, Pilote L, Abrahamowicz M, Marelli AJ. Children and adults with congenital heart disease lost to follow-up: who and when? Circulation 2009;120:302-309.
(27) Beauchesne LM, Therrien J, Alvarez N, Bergin L, Burggraf G, Chetaille P et al. Structure and process measures of quality of care in adult congenital heart disease patients: a pan-Canadian study. Int J Cardiol 2010;157(1):70-74.
(28) Yeung E, Kay J, Roosevelt GE, Brandon M, Yetman AT. Lapse of care as a predictor for morbidity in adults with congenital heart disease. Int J Cardiol 2008;125(1):62-65.
(29) Warnes CA, Liberthson R, Danielson GK, Dore A, Harris L, Hoffman JIE et al. Task Force 1: The changing profile of congenital heart disease in adult life. J Am Coll Cardiol 2001;37(5):1170-1175.
(30) Mackie AS, Rempel GR, Rankin KN, Nicolas D, Magill-Evans J. Risk factors for loss to follow-up among children and young adults with congenital heart disease. Cardiol Young 2012;22(3):307-315.

(31) Reid GJ, Irvine MJ, McCrindle BW, Sananes R, Ritvo PG, Siu SC et al. Prevalence and correlates of successful transfer from pediatric to adult health care among a cohort of young adults with complex congenital heart defects. Pediatrics 2004;113(3 Pt 1):e197-e205.
(32) Iversen K, Vejlstrup NG, Sondergaard L, Nielsen OW. Screening of adults with congenital cardiac disease lost to follow-up. Cardiol Young 2007;17:601-608.
(33) Kaufman M. Role of adolescent development in the transition process. Prog Transplant 2006;16(4):286-290.
(34) Christie D, Viner R. Adolescent development. BMJ 2005;330(7486):301-306.
(35) Stam H, Hartman EE, Deurloo JA, Groothoff J, Grootenhuis MA. Young adult patients with a history of pediatric disease: impact on course of life and transition into adulthood. J Adolesc Health 2006;39(1):4-13.
(36) Lyon ME, Kuehl K, McCarter R. Transition into adulthood in congenital heart disease: missed milestones. J Adolesc Health 2006;39(1):121-124.
(37) Steinberg L. A social neuroscience perspective on adolescent risk-taking. Dev Rev 2008;28(1):78-106.
(38) Louis-Jacques J, Samples C. Caring for teens with chronic illness: risky business? Curr Opin Pediatr 2011;23(4):367-372.
(39) Wickman ME, Anderson NL, Greenberg CS. The adolescent perception of invincibility and its influence on teen acceptance of health promotion strategies. J Pediatr Nurs 2008;23(6):460-468.
(40) Reid GJ, Webb GD, McCrindle BW, Irvine MJ, Siu SC. Health behaviors among adolescents and young adults with congenital heart disease. Congenit Heart Dis 2008;3(1):16-25.
(41) Jalkut MK, Allen PJ. Transition from pediatric to adult health care for adolescents with congenital heart disease: a review of the literature and clinical implications. Ped Nurs 2009;35(6):381-387.
(42) Saidi A, Kovacs AH. Developing a Transition Program from Pediatric- to Adult-Focused Cardiology Care: Practical Considerations. Congenit Heart Dis 2009;4(4):204-215.
(43) van Staa A, van der Stege HA, Jedeloo S, Moll HA, Hilberink SR. Readiness to Transfer to Adult Care of Adolescents with Chronic Conditions: Exploration of Associated Factors. J Adolesc Health 2011;48(3):295-302.
(44) While A, Forbes A, Ullman R, Lewis S, Mathes L, Griffiths P. Good practices that address continuity during transition from child to adult care:synthesis of the evidence. Child Care Healt Dev 2004;30(5):439-452.
(45) Binks JA, Barden WS, Burke TA, Young NL. What do we really know about the transition to adult-centered health care? A focus on cerebral palsy and spina bifida. Arch Phys Med Rehabil 2007;88(8):1064-1073.
(46) Knauth A, Verstappen A, Reiss J, Webb GD. Transition and transfer from pediatric to adult care of the young adult with complex congenital heart disease. Cardiol Clin 2006;24(4):619-629.
(47) Chessa M, De Rosa G, Pardeo M, Negura GD, Butera G, Feslova V et al. Illness understanding in adults with congenital heart disease. Ital Heart J 2005;6(11):895-899.
(48) Van Deyk K, Pelgrims E, Troost E, Goossens E, Budts W, Gewillig M et al. Adolescents' understanding of their congenital heart disease on transfer to adult-focused care. Am J Cardiol 2010;106(12):1803-1807.
(49) Veldtman GR, Matley SL, Kendall L, Quirk J, Gibbs JL, Parsons JM et al. Illness understanding in children and adolescents with heart disease. Heart 2000;84(4):395-397.

(50) Cetta F, Warnes CA. Adults with congenital heart disease: patient knowledge of endocarditis prophylaxis. Mayo Clin Proc 1995;70(1):50-54.
(51) Cetta F, Podlecki DC, Bell TJ. Adolescent knowledge of bacterial endocarditis prophylaxis. J Adolesc Health 1993;14(7):540-542.
(52) Ferencz C, Wiegmann FLJr, Dunning RE. Medical knowledge of young persons with heart disease. J Sch Health 1980;50(3):133-136.
(53) Kantoch MJ, Collins-Nakai RL, Medwid S, Ungstad E, Taylor DA. Adult patients' knowledge about their congenital heart disease. Can J Cardiol 1997;13(7):641-645.
(54) Lesch W, Specht K, Lux A, Frey M, Utens E, Bauer U. Disease-related knowledge and information preferences of young patients with congenital heart disease. Cardiol Young 2014;24(2):321-330.
(55) Moons P, De Volder E, Budts W, De Geest S, Elen J, Waeytens K et al. What do adult patients with congenital heart disease know about their heart disease, treatment, and prevention of complications? Heart 2001;86(1):74-80.
(56) van Engelen K, Baars MJH, van Rongen LT, van der Velde ET, Mulder BJM, Smets EMA. Adults with congenital heart disease: Patients' knowledge and concerns about inheritance. Am J Med Genet Part A 2011;155:1661-1667.
(57) Rönning H, Nielsen NK, Strömberg A, Thilen U, Swahn E. Development and evaluation of a computer-based educational program for adults with congenitally malformed hearts. Eur J Cardiovasc Nurs 2012; 12(1):78-86.
(58) Rönning H, Nielsen NK, Swahn E, Strömberg A. Description and initial evaluation of an educational and psychosocial support model for adults with congenitally malformed hearts. Patient Educ Couns 2011;83:247-251.
(59) Van Deyk K, Moons P, Farroni N, Dedroog D, Pinxten S, Troost E. Effects of a structured education programme for adult patients with congenital heart disease: a follow-up study. Acta Cardiol 2008;63(1):125-126.
(60) McDonagh JE, Southwood TR, Shaw KL. The impact of a coordinated transitional care programme on adolescents with juvenile idiopathic arthritis. Rheumatology (Oxford) 2007;46(1):161-168.
(61) McDonagh JE. Transition of care from paediatric to adult rheumatology. Arch Dis Child 2007;92(9):802-807.
(62) McDonagh JE, Southwood TR, Shaw KL. The impact of coordinated transitional care programme on adolescents with juvenile idiopathic arthritis. Rheumatology 2007;46:161-168.

2 TRANSFER AND TRANSITION AS RESEARCH PRIORITIES IN ACHD NURSING

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Abstract

Background: Since the population of adults with congenital heart disease (CHD) is growing, the role of nurse specialists is expanding. In order to advance ACHD nursing, the establishment of an international nursing research agenda is recommended. We aimed to investigate research priorities as perceived by nurse specialists and researchers in ACHD.

Methods: We applied a sequential quan-qual design. In the quantitative phase, a two-round Delphi study was conducted, in which 37 nurse specialists and nurse researchers in ACHD care participated. Respondents assessed the level of priority of 21 research topics using a 9-point rating scale (1=no priority at all; 9=very high priority). In the qualitative phase, semi-structured interviews were performed with six selected Delphi panelists, to scrutinize pending research questions.

Results: This study revealed that priority should be given to studies investigating knowledge and education of patients, outcomes of Advanced Practice Nursing, quality of life, transfer and transition, and illness experiences and psychosocial issues in adults with CHD. A low priority was given to post-operative pain, sexual functioning, transplantation in ACHD, and health care costs and utilization. Agreement about the level of priority was obtained for 14 out of 21 research topics.

Conclusion: Based on this study, we could develop an international research agenda for ACHD. Researchers ought to focus on these areas of highest priority, in order to expand and strengthen the body of knowledge in ACHD nursing.

Introduction

The number of adults with congenital heart disease (CHD) in society is continuously increasing (1). Based on a population study in Québec, Canada, it is estimated that the prevalence of CHD is 4.09 per 1000 adults (2). Extrapolating these figures to the approximated population of adult individuals (≥ 18 years), there are about 1 million and 2.4 million adult patients with CHD in the USA and in Europe, respectively.

Irrespective of the treatment received in early childhood, patients with CHD remain susceptible to develop medical complications, such as arrhythmias, ventricular dysfunction, endocarditis, and secondary pulmonary hypertension (1,3). They are also prone to experience psychosocial and behavioral problems (4,5) such as anxiety (5), depression (5), compromising health behavior (6) and declined social integration due to problems with employability and insurability (7-10). Specific issues such as reproduction and pregnancy, requires an increasing attention (11,12). Hence, life-long specialized care is recommended for most patients, to prevent and treat possible medical and non-medical problems (13,14). Furthermore, a significant proportion of patients is at risk for re-operation and thus need to be seen regularly in ACHD regional centers and followed for life (15,16). As a result, a series of task forces, consensus statements, and guidelines have been convened to better define the appropriate delivery of care, the practice and providers of care for this growing population of adult patients (15-21). Nursing has played a pivotal role in patient care, teaching and research since adult congenital heart disease (ACHD) programs were first described (22).

According to consensus documents produced by American, Canadian and European societies, nurses who are trained and educated in adult congenital heart disease (ACHD) care, play an integral role of the interdisciplinary ACHD teams (18,20,23-27). Nurse specialists play a key role in assessing patients' needs, counseling patients and their families, and facilitating the transition and transfer of adolescents with CHD to adult care (4,13,22,27,28). Despite the acknowledged role ACHD nurses have made, the nursing aspects of care provided by ACHD nurse specialists is, however, not based on scientific evidence. Therefore, the need to develop a nursing research agenda in ACHD is pivotal to expand the current body of knowledge and furthermore to provide evidence-based ACHD nursing care (4,29). The primary focus of research conducted by nurses includes issues such as quality of life (30-38), illness experiences (39-47), knowledge and education (48-54), and transition and transfer to adult services (55-58).

To further advance the practice of ACHD nursing based on scientific merit, the development of an international nursing research agenda has been recommended (59). The purpose, of this study was to identify research priorities as perceived by nurse specialists and researchers in ACHD nursing.

Materials and methods

Study design

We used a sequential quan-qual design. This is a form of mixed methods research design, comprised of a quantitative (quan) phase, followed by a qualitative (qual) phase (60). In the quantitative phase, a Delphi study was conducted. A Delphi study is a structured consensus method that uses a group facilitation technique, consisting of multiple survey rounds. This technique is used when the goal is to transform the opinions of individual experts into a single group consensus (61-63). A panel of informed individuals, knowledgeable and experienced with the subject under investigation, is identified and compiled (61). These experts are sent a series of multiple structured questionnaires. Hence, the process of a Delphi study is multistaged. Results from each round are reported descriptively to the participants in subsequent rounds. Presentation of the collective opinion provides the participant with additional information, through which the panelist can confirm or adjust the scores given in the preceding survey rounds. Data collection is completed when a predetermined level of consensus is reached within the participating panel (61). In the present study, we used the Delphi method to quantitatively assess the level of priority that ACHD nurse experts give to a set of predefined research topics. For the present study, the Delphi study consisted of two rounds in which electronic questionnaires were individually filled out by the participants.

Upon completion of the Delphi study, we initiated the qualitative phase. Semi-structured telephone interviews were conducted with panelists who participated in both proceeding rounds of the Delphi study.

Sample

Potential participants for the Delphi survey were sought from the membership list of the International Society for Adult Congenital Heart Disease (ISACHD) Nursing Network (64). In addition, names of nursing participants at the 2007 ACHD congress in Philadelphia and personal contacts known

to the authors were added. Finally, the clinic directory of the Adult Congenital Heart Association (www.achaheart.org) was screened to identify hospitals that had nurse specialists in their ACHD program. An email was sent to this list of individuals to identify potential participants. Eligibility criteria were (i) nurses whose clinical practice primarily focused on ACHD care; (ii) held a position as a nurse specialist, nurse practitioner or nurse researcher in ACHD; (iii) expressed their willingness to participate in this Delphi study; and (iv) availability of valid contact information. A total of 47 nurses met the inclusion criteria. During the first Delphi round, 37 of the 47 eligible participants completed the survey form (response rate=78.7%). In the second round, 33 of the 37 nurses who participated in the first round, completed the form (response rate=89.2%). The characteristics of the participants (n=37) are described in **Table 2.1**.

Potential participants for the qualitative phase were selected based on the priority scores they individually gave to predetermined research topics during the Delphi study. Individual ratings of the level of priority were compared to the overall group rating. Potential interviewees were selected if their individual ratings were either divergent or convergent with the overall group rating. Furthermore, potential interviewees were included when they were actively involved in nursing research. A total of six out of eight potential interviewees (75%) agreed to participate.

Table 2.1: Professional characteristics of panelists of the Delphi study (n=37)

Variables	n (%)
Gender	
Female	35 (94.6%)
Male	2 (5.4%)
Age in years (median;Q1-Q3)	44;39-50
Highest level of education	
Certificate in nursing/ Associate degree	5 (13.5%)
Bachelor in nursing/ Polytechnic	4 (10.8%)
Master of Science in Nursing	23 (62.2%)
PhD-student	2 (5.4%)
PhD/Doctorate	3 (8.1%)
Type of hospital	
Local hospital	3 (8.1%)
Regional hospital	1 (2.7%)
University hospital	32 (86.5%)
Other	1 (2.7%)
Work setting (>1 setting possible)	
Outpatient clinic	31 (83.8%)
Cardiac ward/Cardiac surgery	14 (37.8%)
Research/Academic	9 (24.3%)
Education	8 (21.6%)
Other	6 (16.2%)
CCU/ICU	4 (10.8%)
Cathlab	3 (8.1%)
Involvement in nursing research	27 (73.0%)
Years of experience in ACHD (median; Q1-Q3)	5; 3-10
Country	
USA	19 (51.4%)
UK	4 (10.8%)
Canada	4 (10.8%)
Belgium	3 (8.1%)
The Netherlands	3 (8.1%)
Singapore	2 (5.4%)
Denmark	1 (2.7%)
Sweden	1 (2.7%)

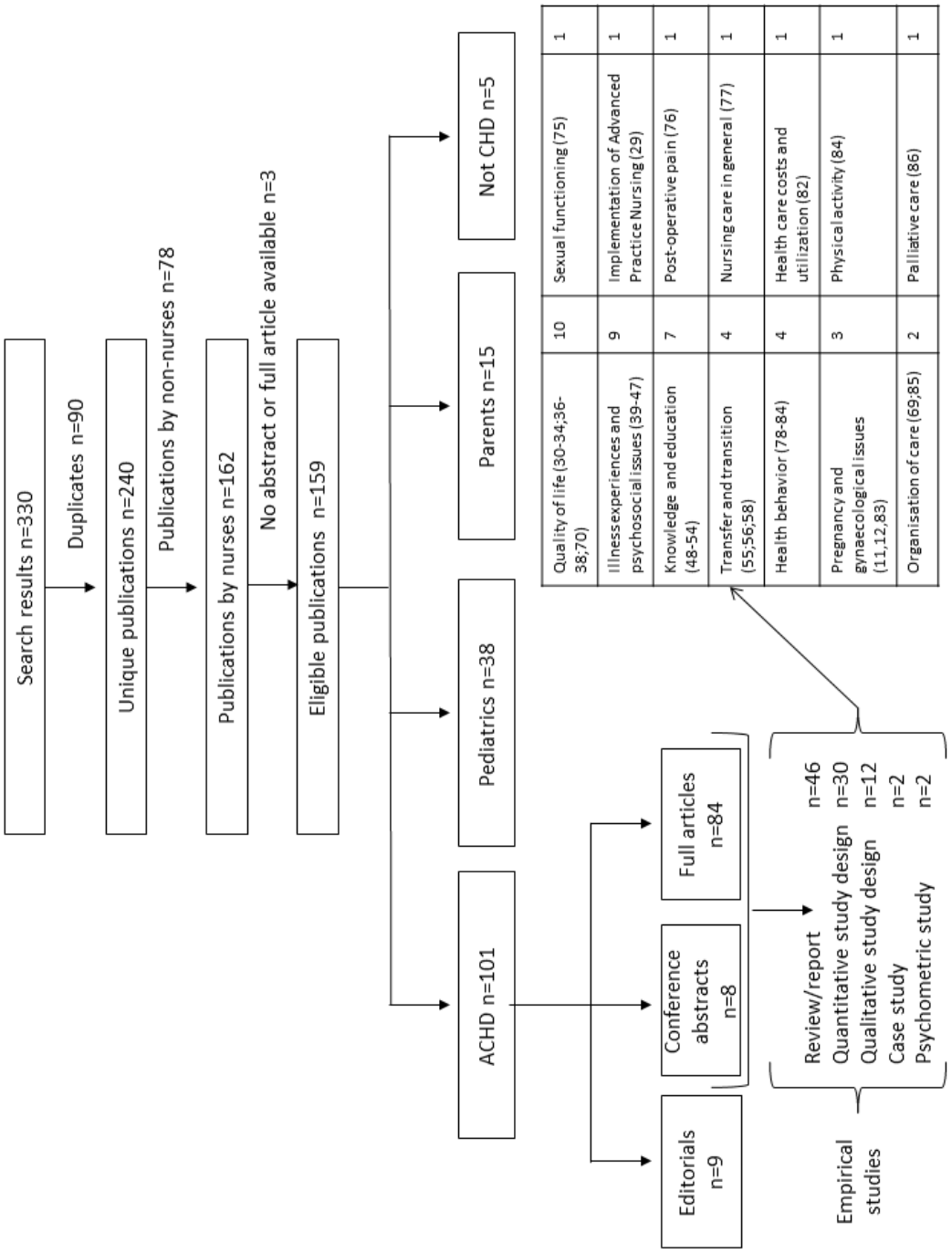
Delphi survey instrument

The Delphi survey form was developed in two stages. First, a literature review was performed to determine the topics already investigated in nurse-led studies. We conducted a systematic literature search in the databases Medline and Cinahl, using the keywords 'congenital heart' AND adult AND nurs*. This search resulted in 330 hits (**Figure 2.1**), published between January 1980 and July 2010.

After elimination of duplicates, publications by non-nurses, and articles for which no abstract or full text was available, 159 publications were identified for further analysis. Overall, 101 articles pertained to ACHD, nine of which were editorials, eight were conference abstracts, and 84 were full articles. Forty-six articles were either reviews or reports, leaving 64 empirical studies for systematic analysis. The majority of these empirical nursing studies investigated quality of life (n=10) (30-38;62), followed by illness experiences and psychosocial issues (n=9) (39-47), knowledge and education (n=7) (48-54), transfer and transition (n=4) (55-58), and health behavior (n=4) (66-69) (**Figure 2.1**). Issues that were investigated to a lesser extent were: pregnancy and gynaecological issues (11,12,70); organization of care (71,72); sexual functioning (73); implementation of advanced practice nursing (29); post-operative pain (74); nursing care in general (75); health care costs and utilization (76); physical activity (77); and palliative care (78). Based on the results of this systematic literature review, a preliminary list of 13 research topics in ACHD nursing was composed.

This preliminary list was then presented to the executive board of the ISACHD Nursing Network. The members of this board were asked to propose additional topics for nursing research. Eight additional topics were proposed, resulting in a total of 21 ACHD-related research topics. These 21 topics were included in the final survey form for round one of the Delphi study. Respondents were asked to indicate the level of priority for each of the 21 predefined research topics, using a 9 point-rating scale (1=no priority at all; 9=very high priority). Free text space was provided, giving participants the opportunity to generate ideas and suggest additional research topics based on their professional experiences. The survey form was accompanied by a short self-report questionnaire on professional characteristics.

Figure 2.1: Flowchart systematic literature review



Data collection

Eligible experts received an electronic invitation explaining the aims of the study and describing the methodological principles of a Delphi survey. A questionnaire was attached to this letter with instructions to be completed within two weeks. Non-responders received up to three reminders.

After data collection was completed in round 1, data were analyzed descriptively (median, Q1-Q3 and range).The panelists were then invited to partake in round two. In this second round, all panelists were given their personal responses from round one, the overall group rating of all 21 predefined research topics. Furthermore, panelists were given the opportunity to reconsider their initial scores in light of the results of the preceding round. Again, in the second Delphi round, experts indicated the level of priority of each research topic using a 9-point rating scale. Data collection in round two was completed when there was agreement for more than two-thirds of the 21 research topics.

Next, to further scrutinize the pending research questions in depth, qualitative data collection commenced. Using an interview guide, semi-structured telephone interviews, were conducted. Individual interview guides were developed using the individual ratings given to the research topics in the quantitative phase. Interviewees were asked to clarify the priority levels they assigned to certain research topics. Furthermore, interviewees were asked to formulate research questions they would suggest for future studies regarding each of the research topics in the top five. Six interviews, each lasting 30-60 minutes, were conducted audio taped and transcribed verbatim.

Data analysis

Descriptive statistics of the quantitative data from the two sequential Delphi rounds were obtained using Statistical Package for the Social Sciences 17.0 (SPSS Inc., Chicago, IL). Frequency tables of attributed priority scores were composed and median scores, range of scores and quartiles (Q1-Q3) were calculated. Furthermore, priority scores collected in round two of the Delphi study were analyzed using RIDIT analysis in order to classify the research topics according to an increasing level of priority. RIDIT analysis is a statistically valid method to analyze ordinal data (79,80). In this analysis technique, the distribution of item scores is compared with a reference distribution. We used the distribution of priority scores over all research topics in our sample of panelists as the reference distribution. The RIDIT obtained for each item reflects the probability that this particular research topic received a higher priority score than another randomly selected topic of the list (81).

RIDIT's value ranges from 0 to 1, and higher RIDIT-scores indicate a greater chance that a specific research topic received a higher priority than another randomly selected topic.

Additionally, a Disagreement Index (DI) was calculated for each research topic, to evaluate whether consensus was reached about the assigned level of priority. The DI expresses the diffusion of ratings. To calculate the DI, the Interpercentile Range (IPR) is divided by the Interpercentile Range Adjusted for Symmetry (IPRAS). A DI-value greater than 1 indicates significant disagreement (82).

Finally, qualitative data obtained from the semi-structured interviews were analyzed using a descriptive content analysis, in which verbatim transcripts were labeled.

Results

Quantitative analysis of research priorities in ACHD nursing

Analysis of the priority scores revealed that 17 out of 21 ACHD-related research topics received a median score ≥ 7 on a scale from 1 to 9 (**Figure 2.2**). The four remaining research topics received a median priority score of five or six. Analysis of the range of scores for each research topic, revealed a great variability in the lowest score, whereas the highest score was always nine, with the exception of one topic that received a maximum score of eight.

Based on RIDIT-analysis, we were able to determine a rank order of research priorities (**Figure 2.2**). The research topics that were given top priority in ACHD nursing were knowledge and education of patients, followed by outcomes of advanced practice nursing (APN), quality of life, transfer and transition to ACHD care, illness experiences and psychosocial issues, pregnancy and gynecological issues, health behavior, exercise capacity, role development of APN, organization of care, and palliative care and end-of-life issues. A low priority was given to post-operative pain, sexual functioning, transplantation in ACHD, and health care costs and utilization.

Analysis of the Disagreement Index (DI) demonstrated that there was agreement on the priority scores given to 14 out of 21 (66.7%) suggested topics. There was agreement on the 13 highest ranked research topics, as well as on the lowest ranked topic. There was disagreement among the panel of experts on the priority that should be assigned to the topics regarding long-term survival, survival outcomes of specific interventions, cognition issues, nursing care in general, health care costs and utilization, transplantation in ACHD and sexual functioning.

Qualitative analysis of research priorities in ACHD nursing

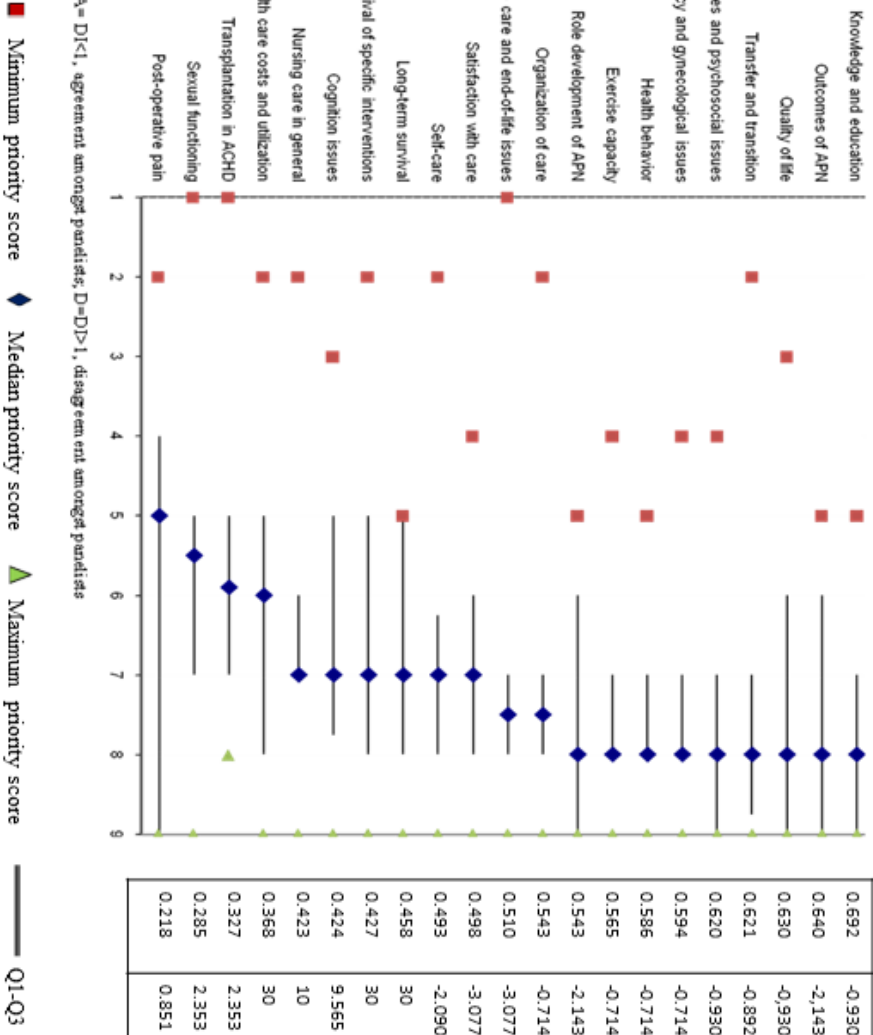
In semi-structured interviews, participants were asked to reflect on specific themes and research questions for the five topics that received the highest priority in the Delphi study. The themes and research questions suggested are listed in **Table 2.2**. For knowledge and education of patients, specific research questions were formulated with respect to (i) patients' knowledge of the impact of the disease; (ii) development of an educational plan for life; and (iii) learning difficulties or neuro-cognitive impairment in patients with CHD. Research on outcomes of the APN role should specifically target the implementation of different APN roles in ACHD care. For quality-of-life research, it is suggested to undertake studies in specific subgroups of patients with CHD. Furthermore, studies on the development of a transition program for adolescents with CHD, and evaluation of the effectiveness of different models of transitional care are advocated (**Table 2.2**).

Discussion

ACHD is a growing area of practice, one in which a growing number of nurses are directly involved. Numerous ACHD programs have nurse specialists on staff (71) and are assumed to provide evidence-based care. In addition, an increasing number of nurses are undertaking empirical studies in adults with CHD. As providers are preparing for the next decade of ACHD care, it is important to know the priority of topics to be examined in future studies. Therefore, in this study we examined the research priorities as perceived by nurse specialists and nurse researchers in ACHD and, subsequently, developed an international nursing research agenda. The five most important areas for ACHD nursing research were: patient's knowledge and education, APN outcomes, quality of life, transfer and transition to adult CHD care, and illness experiences and psychosocial issues.

Indeed, highest priority was given to studies investigating the knowledge and education of adults with CHD. As pediatric patients move into adult setting, it is assumed they are prepared to become responsible for their life and health care. In order to take up this responsibility, however, adults with CHD need to have sufficient knowledge about their disease, treatment and preventive measures. To date, seven studies were conducted by nurses concerning knowledge and education (48-54). These studies have reported that important gaps in the knowledge of these patients exist concerning their heart defect, treatment and preventive measures; and pointed out that there is a need to develop, implement and evaluate structured educational activities (52,54).

Figure 2.2: Priority scores of 21 predefined research topics in ACHD



	Ridit	DI	Rating[1]
Knowledge and education	0.692	-0.930	A
Outcomes of APN	0.640	-2,143	A
Quality of life	0.630	-0,930	A
Transfer and transition	0.621	-0.892	A
Illness experiences and psychosocial issues	0.620	-0.930	A
Pregnancy and gynecological issues	0.594	-0.714	A
Health behavior	0.586	-0.714	A
Exercise capacity	0.565	-0.714	A
Role development of APN	0.543	-2.143	A
Organization of care	0.543	-0.714	A
Palliative care and end-of-life issues	0.510	-3.077	A
Satisfaction with care	0.498	-3.077	A
Self-care	0.493	-2.090	A
Long-term survival	0.458	30	D
Survival of specific interventions	0.427	30	D
Cognition issues	0.424	9.565	D
Nursing care in general	0.423	10	D
Health care costs and utilization	0.368	30	D
Transplantation in ACHD	0.327	2.353	D
Sexual functioning	0.285	2.353	D
Post-operative pain	0.218	0.851	A

Table 2.2: Suggested research questions on top 5 research priorities in ACHD (n=6)

1. KNOWLEDGE AND EDUCATION OF PATIENTS WITH CHD

(i) Knowledge on the impact of the disease:

- What is the knowledge of patients on continuity of care (e.g., knowledge on health care insurance, reasoning for follow-up, recommended frequency of follow-up visits, how to navigate through the healthcare system)?
- What is the knowledge of patients regarding pharmacological treatment (e.g., prescribed frequency and doses of medication, possible side-effects, consequences of non-adherence)?
- What is the knowledge of patients, both sexes, on pregnancy counseling (e.g., knowledge on potential risks and complications during pregnancy)?
- What is the knowledge of patients concerning recommended and/or allowed physical activities with regard to their heart condition?

(ii) Development of an educational plan for life:

- What do patients want to know at different stage of their life and their disease?
- Which components from educational models implemented in patients with a chronic condition can be implemented in CHD care?
- Which educational interventions or programs are effective in enhancing the level of knowledge in patients with CHD?

(iii) Learning difficulties and neuro-cognitive impairment:

- What is the proportion of patients with CHD who are neuro-cognitively impaired?
- Which factors could predict the development of learning difficulties in patients with CHD?

2. OUTCOMES OF ADVANCED PRACTICE NURSING

(i) Evaluation of the implementation of APN roles:

- Which outcomes, parameters and/or indicators should be measured in order to evaluate the impact of different APN roles?
- What is the impact of APN roles in ACHD care on the predefined outcomes, parameters and/or indicators related?

3. QUALITY OF LIFE

- What is the quality of life in the overall sample of patients with CHD?
- What is the quality of life in specific subgroups of patients with CHD (e.g., cyanotic heart lesions) based on their clinical characteristics?
- What is the quality of in children and adolescents with CHD?
- What is the quality of life in adult patients with CHD?
- What is the quality of life in patients with CHD across different levels of care and settings for cardiac follow-up?
- What are the psychometric properties of the instruments used to investigate quality of life?

4. TRANSFER AND TRANSITION TO ADULT HEALTH SERVICES

(i) Development of a transition program:

- What is the core content or the key elements of a transition program for patients with CHD?

(ii) Evaluation of the effectiveness of transition programs:

- Which outcomes, parameters and/or indicators should be measured in order to evaluate the effectiveness of a transition program in patients with CHD?
- Which factors influence continuity of care in adults with CHD?
- What benefits could a transition program give to patients, parents, healthcare workers and the healthcare system in CHD care?
- What is the effectiveness of different organizational models of transition and transfer of care in patients with CHD?

5. ILLNESS EXPERIENCES AND PSYCHOSOCIAL ISSUES

- What is the impact of illness experiences of patients with CHD on their daily living?
- Which psychosocial and behavioral problems occur in adolescents with CHD (e.g., anxiety, depression, etc.)?

In the present study, participants indicated that priority should be given to studies assessing the level of knowledge of patients regarding the need for follow-up, their treatment, pregnancy counseling and physical activities. Further, future research should focus on the development and evaluation of an educational plan for the lifespan of patients.

In our study, the second research priority was research on the outcomes of APN. Indeed, when performing our systematic review, there were no publications identified which investigated the outcomes of the implementation of the APN role. However, to advance the role of ACHD nursing, it is important to measure the impact of APN and to identify targets for improvement. Although during our Delphi study, a lower priority score was assigned to studies examining role development of APN, interviewees stressed however the importance of conducting research on the development and impact of the APN role. Internationally, a number of different roles are defined within the scope of APN. To better define the qualifications, tasks, responsibilities and outcomes of ACHD APNs, future research on role development is needed.

The third priority in the research agenda was quality of life in adults with CHD. To date, ten nurse led studies were conducted on quality of life in ACHD patients (30-38;65). Hence, quality of life is the topic most frequently investigated by nurses involved in ACHD care. Furthermore, a survey among European ACHD nurse specialists showed that 90% of the nurse specialists who were involved in nursing research, conducted quality-of-life research (29). Therefore, it could be counterintuitive that quality-of-life research is still highly needed. The present study, however, revealed that additional studies on quality of life are required, particularly studies conducted in specific subgroups of patients based on for example their age, type of heart defect or other clinical characteristics. Additionally, it is for example, important to identify patients with a high risk for poor quality of life, and investigate interventions to improve their quality of life.

The fourth priority was the transition and transfer of adolescents with CHD to adult-focused facilities. Since the majority of patients with CHD need cardiac follow-up throughout their entire life span (16,18,20,83-85), a timely transfer of care from pediatric cardiology to different levels of adult-focused care has been recommended (18-20;85). Several consensus documents have described the need for the implementation of transition programs that prepare adolescents with CHD for the transfer of care when becoming an adult (18,27,85). To date, only four nurse-led studies in this area were found. These studies either investigated the experiences and expectations of patients with CHD regarding transfer and transition (56,58), or studied the current practices on transfer and transition

in 69 European and North-American centers (55). The need for additional research on the development of transition programs, subsequently followed by experimental studies evaluating the effectiveness of such programs, was expressed in our study.

The fifth research priority was the illness experiences and psychosocial issues of patients with CHD. In order to give patients comprehensive care, attention must be given to both medical and non-medical complications patients might experience (4). Nurses need to comprehend which psychosocial obstacles patients face when growing up with CHD. It is suggested that future research should focus on the impact of illness experiences of adults with CHD in their daily living.

This study was the first to establish an international nursing research agenda for ACHD nursing. However, there are some limitations to bear in mind when interpreting the results of this study. First, only 73% of the panelists who participated in the Delphi study were involved in research. This could imply that nurses gave priority to certain research topics without taking the feasibility of such studies into account. Second, when using the Delphi technique, consideration must be given to the level of consensus to be employed. However, a universally recommended level of agreement does not exist. McKenna et al. suggested a minimal agreement of 51% amongst panelists, while others would recommend to obtain a consensus level of 65%, 70% or even 80% (63,86). We chose to terminate data collection through the subsequent Delphi rounds, when agreement on the priority scores was obtained for at least two third of the 21 research topics. We calculated a Disagreement Index for each of the 21 predefined research topics in order to evaluate whether agreement was obtained. Third, because we used a mixed methods design, we had the opportunity to investigate the top five research priorities in depth by conducting semi-structured interviews. These interviews revealed that panelists possibly indicated certain research priorities based on their own hospital experience and current working setting suggesting possible bias. For example, when a nurse worked in a hospital where patients were not transferred from pediatric cardiology to an adult-focused facility because the hospital provides one comprehensive program for the entire life-span of the patient, nurses gave a low priority score to this research area.

Conclusion

Based on this mixed methods study, an international nursing research agenda on ACHD has been established. Topics that should receive the highest priority are: patient knowledge and education, APN outcomes, quality of life, transfer and transition to adult care, and illness experiences and psychosocial issues. Hence, current and future nurse researchers should consider these areas in

order to expand the evidence basis, strengthen the body of knowledge and prepare ACHD nursing for the next decade.

References

1. Brickner ME, Hillis LD, Lange RA. Congenital heart disease in adults. First of two parts. N Engl J Med 2000;342:256-263.
2. Marelli AJ, Mackie AS, Ionescu-Ittu R, Rahme E, Pilote L. Congenital heart disease in the general population: changing prevalence and age distribution. Circulation 2007;115:163-172.
3. Brickner ME, Hillis LD, Lange RA. Congenital heart disease in adults. Second of two parts. N Engl J Med 2000;342:334-342.
4. Moons P, De Geest S, Budts W. Comprehensive care for adults with congenital heart disease: expanding roles for nurses. Eur J Cardiovasc Nurs 2002;1:23-28.
5. Karsdorp PA, Everaerd W, Kindt M, Mulder BJM. Psychological and cognitive functioning in children and adolescents with congenital heart disease: a meta-analysis. J Ped Psychol 2007;32:527-541.
6. Reid GJ, Webb GD, McCrindle BW, Irvine MJ, Siu SC. Health behaviors among adolescents and young adults with congenital heart disease. Congenit Heart Dis 2008;3:16-25.
7. Hart EM, Garson A. Psychosocial concerns of adults with congenital heart disease. Employability and insurability. Cardiol Clin 1993;11:711-715.
8. Kokkonen J, Paavilainen T. Social adaptation of young adults with congenital heart disease. Int J Cardiol 1992;36:23-29.
9. Kaemmerer H, Tintner H, Konig U, Fritsch J, Sechtem U, Hopp HW. Psychosocial problems of adolescents and adults with congenital heart defects. Z Kardiol 1994;83:194-200.
10. McGrath KA, Truesdell SC. Employability and career counseling for adolescents and adults with congenital heart disease. Nurs Clin North Am 1994;29:319-330.
11. Canobbio MM, Mair DD, van der Velde M, Koos BJ. Pregnancy outcomes after the Fontan repair. J Am Coll Cardiol 1996;28:763-767.
12. Canobbio MM, Mair DD, Rapkin AJ, Perloff JK, George BL. Menstrual patterns in females after the Fontan repair. Am J Cardiol 1990;66:238-240.
13. Perloff JK, Miner PD. Specialized facilities for the comprehensive care of adults with congenital heart disease. In: Perloff JK, Child JS, eds. Congenital heart disease in adults. Philadelphia: W.B. Saunders Company, 1998 p.9-12.
14. Child JS, Collins-Nakai RL, Alpert JS, et al. Task force 3: workforce description and educational requirements for the care of adults with congenital heart disease. J Am Coll Cardiol 2001;37:1183-1187.
15. Warnes CA, Liberthson R, Danielson GK, et al. Task force 1: the changing profile of congenital heart disease in adult life. J Am Coll Cardiol 2001;37:1170-1175.
16. Landzberg MJ, Murphy DJ Jr., Davidson WR Jr., et al. Task force 4: organization of delivery systems for adults with congenital heart disease. J Am Coll Cardiol 2001;37:1187-1193.
17. Sable C, Foster E, Uzark K, et al. Best Practices in Managing Transition to Adulthood for Adolescents With Congenital Heart Disease: The Transition Process and Medical and Psychosocial Issues: A Scientific Statement From the American Heart Association. Circulation 2011; 123(13):1454-1485.

18. Deanfield J, Thaulow E, Warnes C, et al. Management of grown up congenital heart disease. Eur Heart J 2003;24:1035-1084.
19. Foster E, Graham TP Jr., Driscoll DJ, et al. Task force 2: special health care needs of adults with congenital heart disease. J Am Coll Cardiol 2001;37:1176-1183.
20. Warnes CA, Williams RG, Bashore TM, et al. ACC/AHA 2008 Guidelines for the Management of Adults with Congenital Heart Disease: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (writing committee to develop guidelines on the management of adults with congenital heart disease). Circulation 2008;118:e714-e833.
21. Baumgartner H, Bonhoeffer P, De Groot NM, et al. ESC Guidelines for the management of grown-up congenital heart disease (new version 2010). Eur Heart J 2010;31:2915-2957.
22. Perloff JK. The UCLA Adult Congenital Heart Disease Program. Am J Cardiol 1986;57:1190-1192.
23. Therrien J, Dore A, Gersony W, et al. CCS Consensus Conference 2001 update: recommendations for the management of adults with congenital heart disease. Part I. Can J Cardiol 2001;17:940-959.
24. Fifth report on the provision of services for patients with heart disease. Heart 2002;88 Suppl 3:iii1-iii56.
25. Report of the paediatric and congenital cardiac services review group. London: Department of Health; 2003. p. 1-49.
26. Congenital heart disease after childhood: an expanding patient population. 22nd Bethesda Conference, Maryland, October 18-19, 1990. J Am Coll Cardiol 1991;18:311-342.
27. Moodie DS. Adult congenital heart disease. Curr Opin Cardiol 1994;9:137-142.
28. Canobbio MM, Day MJ. The role of the clinical specialist in an adult congenital heart disease program. Nurs Clin North Am 1994;29:357-366.
29. Moons P, Scholte op Reimer W, De Geest S, et al. Nurse specialists in adult congenital heart disease: The current status in Europe. Eur J Cardiovasc Nurs 2006;5:60-67.
30. Moons P, Van Deyk K, De Bleser L, et al. Quality of life and health status in adults with congenital heart disease: a direct comparison with healthy counterparts. Eur J Cardiovasc Prev Rehabil 2006;13:407-413.
31. Moons P, Van Deyk K, Marquet K, et al. Individual quality of life in adults with congenital heart disease: a paradigm shift. Eur Heart J 2005;26:298-307.
32. Moons P, Van Deyk K, De Geest S, Gewillig M, Budts W. Is the severity of congenital heart disease associated with the quality of life and perceived health of adult patients? Heart 2005;91:1193-1198.
33. Simko LC, McGinnis KA. What is the perceived quality of life of adults with congenital heart disease and does it differ by anomaly? J Cardiovasc Nurs 2005;20:206-214.
34. Moons P, De Bleser L, Budts W, et al. Health status, functional abilities and quality of life following the Mustard or Senning operation. Ann Thorac Surg 2004;77:1359-1365.
35. Moons P, Marquet K, Budts W, De Geest S. Validity, reliability and responsiveness of the "Schedule for the Evaluation of Individual Quality of Life - Direct Weighting" (SEIQoL-DW) in congenital heart disease. Health Qual Life Outcomes 2004;2:27.
36. Simko, LC. Quality of life of adults with congenital heart disease. Duquesne University; 2000. p. 1-125.
37. Simko LC. Adults with congenital heart disease: utilizing quality of life and Husted's nursing theory as a conceptual framework. Crit Care Nurs Q 1999;22:1-11.

38. Moons P, Van Deyk K, Marquet K, De Bleser L, De Geest S, Budts W. Profile of adults with congenital heart disease having a good, moderate, or poor quality of life: a cluster analytic study. Eur J Cardiovasc Nurs 2009;8:151-157.
39. Young QR, Ignaszewski A, Fofonoff D, Kaan A. Brief screen to identify 5 of the most common forms of psychosocial distress in cardiac patients: validation of the screening tool for psychological distress. J Cardiovasc Nurs 2007;22:525-534.
40. Balon YE, Then KL, Rankin JA, Fung T. Looking beyond the biophysical realm to optimize health: results of a survey of psychological well-being in adults with congenital cardiac disease. Cardiol Young 2008;18:494-501.
41. Claessens P, Moons P, de Casterle BD, Cannaerts N, Budts W, Gewillig M. What does it mean to live with a congenital heart disease? A qualitative study on the lived experiences of adult patients. Eur J Cardiovasc Nurs 2005;4:3-10.
42. Gantt L. As normal a life as possible: mothers and their daughters with congenital heart disease. Health Care Women Int 2002;23:481-491.
43. Kools S, Tong EM, Hughes R, et al. Hospital experiences of young adults with congenital heart disease: divergence in expectations and dissonance in care. Am J Crit Care 2002;11:115-125.
44. McMurray R, Kendall L, Parsons JM, et al. A life less ordinary: growing up and coping with congenital heart disease. Coronary Health Care 2001; 5:51-7.
45. Tong EM, Sparacino PS, Messias DK, Foote D, Chesla CA, Gilliss CL. Growing up with congenital heart disease: the dilemmas of adolescents and young adults [see comment]. Cardiol Young 1998;8:303-309.
46. Gantt LT. Growing up heartsick: the experiences of young women with congenital heart disease. Health Care Women Int 1992;13:241-248.
47. Aoki, M. The creation of a natural sense: the self-construction of children with congenital heart disease by body-image. Journal of Japan Academy of Nursing Science 2009;3:43-51.
48. Moons P, Fleck D, Jaarsma T, et al., and on behalf of the Undertaking Nursing Interventions Throughout Europe (UNITE) Study Group. What do cardiovascular nurses know about the hematological management of patients with Eisenmenger syndrome? Eur J Cardiovasc Nurs 2009; 8: 246-250.
49. Van Deyk K, Pelgrims E, Troost E, et al. Adolescents' understanding of their congenital heart disease on transfer to adult-focused care. Am J Cardiol 2010;106:1803-1807.
50. Rönning H, Swahn E, Nielsen NE, Strömberg A. Development of a Knowledge Questionnaire for Adults With Congenital Heart Disease [abstr]. Eur J Cardiovasc Nurs 2006;5(S1):S3-S54.
51. Simko LC, McGinnis KA, Schembri J. Educational needs of adults with congenital heart disease. J Cardiovasc Nurs 2006;21:85-94.
52. Van Deyk K, Moons P, Dedroog D, Pinxten S, Troost E, Budts W. Knowledge of Adult Patients With Congenital Heart Disease: 5 Years Later [abstr]. Eur J Cardiovasc Nurs 2006;5: S37.
53. Liljeroos H, Swahn E, Stromberg A, Nielsen NE. Development of a Computer-Based Educational Programme for Adults With Congenital Heart Disease [abstr]. Eur J Cardiovasc Nurs 2005;4:74-75.
54. Moons P, De Volder E, Budts W, et al. What do adult patients with congenital heart disease know about their disease, treatment, and prevention of complications? A call for structured patient education. Heart 2001;86:74-80.

55. Hilderson D, Saidi AS, Van Deyk K, et al. Attitude toward and current practice of transfer and transition of adolescents with congenital heart disease in the United States of America and Europe. Pediatr Cardiol 2009;30:786-793.
56. Moons P, Pinxten S, Dedroog D, et al. Expectations and experiences of adolescents with congenital heart disease on being transferred from pediatric cardiology to an adult congenital heart disease program. J Adolesc Health 2009;44:316-322.
57. Clarizia NA, Chahal N, Manlhiot C, Kilburn J, Redington AN, McCrindle BW. Transition to adult health care for adolescents and young adults with congenital heart disease: perspectives of the patient, parent and health care provider. Can J Cardiol 2009;25:e317-e322.
58. Kools S, Gilliss CL, Tong EM. Family transitions in congenital heart disease management: the impact of hospitalization in early adulthood. J Fam Nurs 1999;5: 404-425.
59. Moons P, Hilderson D, Van Deyk K. Congenital cardiovascular nursing: preparing for the next decade. Cardiol Young 2009;19 (Suppl 2):106-111.
60. Creswell JW, Plano Clark VL. Designing and Conducting Mixed Methods Research. California, USA: SAGE publications, Inc; 2007.
61. Hasson F, Keeney S, McKenna H. Research guidelines for the Delphi survey technique. J Adv Nurs 2000;32:1008-1015.
62. Keeney S, Hasson F, McKenna H. Consulting the oracle: ten lessons from using the Delphi technique in nursing research. J Adv Nurs 2006;53:205-212.
63. Mcilfatrick SJ, Keeney S. Identifying cancer nursing research priorities using the Delphi technique. J Adv Nurs 2003;42:629-636.
64. Moons P, Canobbio MM, Harrison J. The International Adult Congenital Heart Disease Nursing Network: coming together for the future. Prog Cardiovasc Nurs 2006;21:94-96.
65. Simko LC, McGinnis KA. Quality of life experienced by adults with congenital heart disease. AACN Clin Issues 2003;14:42-53.
66. Chen CW, Chen YC, Chen MY, Wang JK, Su WJ, Wang HL. Health-promoting behavior of adolescents with congenital heart disease. J Adolesc Health 2007;41:602-609.
67. Moons P, Van Deyk K, Dedroog D, Troost E, Budts W. Prevalence of cardiovascular risk factors in adults with congenital heart disease. Eur J Cardiovasc Prev Rehabil 2006;13:612-616
68. Ratner PA, Johnson JL, Richardson CG, et al. Efficacy of a smoking-cessation intervention for elective-surgical patients. Res Nurs Health 2004;27:148-161.
69. Moons P, Budts W, Costermans E, Huyghe E, Pieper PG, Drenthen W. Pregnancy-related health behavior of women with congenital heart disease: room for behavioral change interventions. Congenit Heart Dis 2009;4:348-355.
70. Kilpatrick K, Purden M. Using reflective nursing practice to improve care of women with congenital heart disease considering pregnancy. MCN Am J Matern Child Nurs 2007;32:140-147
71. Moons P, Meijboom FJ, Baumgartner H, Trindade PT, Huyghe E, Kaemmerer H. Structure and activities of adult congenital heart disease programmes in Europe. Eur Heart J 2010;31:1305-1310.
72. Moons P, Engelfriet P, Kaemmerer H, Meijboom FJ, Oechslin E, Mulder BJ. Delivery of care for adult patients with congenital heart disease in Europe: results from the Euro Heart Survey. Eur Heart J 2006;27:1324-1330
73. Moons P, Van Deyk K, Marquet K, De Bleser L, Budts W, De Geest S. Sexual functioning and congenital heart disease: Something to worry about? Int J Cardiol 2007;121:30-35.

74. Jensen L, Rebeyka D, Urquhart G, Roschkov S. Pain in adults post-surgical repair of congenital heart defects. Can J Cardiovasc Nurs 2004;14:8-17.
75. Laiti LA, Roselle-Kollauf C. Congenital heart diseases in adults. Nursing grand rounds. J Cardiovasc Nurs 1989;3:74-78.
76. Moons P, Siebens K, De Geest S, Abraham I, Budts W, Gewillig M. A pilot study of expenditures on, and utilization of resources in, health care in adults with congenital heart disease. Cardiol Young 2001;11:301-313.
77. Berg SK, Pedersen PU. Perception of general health in adults with congenital heart disease who no longer attend medical follow-up. Eur J Cardiovasc Nurs 2008;7(4):264-268.
78. Tellett L, Davis C. The case study masterclass. Case 43 answers: Fulfilling a patient's to go home from intensive care. Eur J Palliativ Care 2009;16(3):122.
79. Donaldson GW. Ridit scores for analysis and interpretation of ordinal pain data. European Journal of Pain 1998;2:221-227.
80. Bross, I. How to use ridit analysis. Biometrics 1985;14:18-38.
81. Sermeus W, Delesie L. Ridit analysis on ordinal data. West J Nurs Res 1996;18:351-359.
82. Fitch K. The RAND/UCLA appropriateness Method. User's Manual; 2000. Lazaro P, Aguilar MD, Bernand B, et al.
83. Webb GD, Williams RG. Care of the adult with congenital heart disease: introduction. J Am Coll Cardiol 2001;37:1166.
84. Silversides CK, Marelli A, Beauchesne L, et al. Canadian Cardiovascular Society 2009 Consensus Conference on the management of adults with congenital heart disease: executive summary. Can J Cardiol 2010;26:143-150.
85. Murphy Jr DJ, Foster E. ACCF/AHA/AAP recommendations for training in pediatric cardiology. Task force 6: training in transition of adolescent care and care of the adult with congenital heart disease. J Am Coll Cardiol 2005;46:1399-1401.
86. Sumsion, T. The Delphi technique: an adaptive research tool. Br J Occup Ther 1998;61(4):153-156.

3 TRANSFER DESTINATIONS OF YOUNG PEOPLE LEAVING PEDIATRIC CARDIOLOGY

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Abstract

Objectives: The transfer of adolescents with congenital heart disease from pediatric to adult care was examined. The aims were to investigate where these adolescents received adult-centered care, to determine the proportion of patients with no follow-up and with no appropriate follow-up after leaving pediatric cardiology, and to explore the determinants of no-follow-up and no appropriate follow-up.

Background: Even after successful treatment, many patients require lifelong cardiac surveillance by specialized practitioners. Although guidelines describe the most appropriate level of follow-up, this is not always implemented in practice.

Methods: A descriptive, observational study was performed, including 794 patients with congenital heart disease examined and/or treated at a tertiary care center.

Results: Overall, 58 of the 794 patients included (7.3%) were not in follow-up. Cessation of follow-up was found in 2 of 74 patients with complex (2.7%), 31 of 448 patients with moderate (6.9%), and 25 of 272 patients with simple (9.2%) heart defects, Moreover, 684 patients (86.1%) remained in specialized follow-up. According to international guidelines, 81 patients (10.2%) did not receive the minimal level of cardiac care. Multivariable logistic regression revealed that male sex and no prior heart surgery were associated with no follow-up. Male sex, no prior heart surgery, and greater complexity of congenital heart disease were associated with no appropriate level of cardiac follow-up.

Conclusions: The proportion of patients in this study lost to follow-up was substantially lower than in other Western countries. Because only patient-related factors were examined with respect to loss to follow-up, further examination of patient-related, hospital-related, and healthcare-related determinants of lack of follow-up is needed.

Introduction

Congenital heart disease (CHD) is considered to be the most common birth defect, with an incidence of 0.8% in newborns (1). A substantial increase in life expectancy has been observed in the past decades, with approximately 90% of children born with CHD surviving into adulthood (2). Despite this improvement, patients with CHD can experience residua or sequelae of the initial treatment they received. Therefore, these defects are considered to be "repaired" rather than "cured". Hence, even after successful primary treatment or surgery, many patients with CHD require lifelong cardiac surveillance by specialized practitioners (3-5).

International guidelines and consensus statements describe the most appropriate setting for follow-up of patients with CHD. During childhood, these patients are most appropriately followed up in pediatric programs. As they approach adulthood, a timely transfer to an adult congenital heart disease (ACHD) program is advocated (6-9). However, different levels of adult-centered care exist. For instance, guidelines categorize the level of ACHD care into 3 types: 1) specialist care; (2) shared care; and (3) non-specialist care (6). Specialist care is follow-up given by specialized ACHD cardiologists and is provided mainly at tertiary care centers. Shared care is follow-up given by a general adult cardiologist in close collaboration with a CHD specialist. Nonspecialist care is follow-up given by a general or community cardiologist, or a general practitioner, with access to specialized care if needed (6).

Guidelines describe which level of care is most appropriate for each type of heart defect (5,6,9). Except for patients with ligated and divided ductus arteriosus, all patients should continue to receive cardiac care from a specialized ACHD program, a local healthcare provider, or a collaboration between local and specialist providers (9). Patients with complex heart defects, such as cyanotic heart disease or transposition of the great arteries, should receive checkups every 6 to 12 months at a specialist center (5). Patients with moderate -complexity lesions, such as Tetralogy of Fallot, atrioventricular septal defects, or coarctation of the aorta, should have follow-up visits every 1 to 2 years (5). This is preferably done at specialist centers (5) but can also be undertaken at shared care facilities if the CHD course is uncomplicated (6). Patients with simple heart defects, such as small atrial septal defects or patent ductus arteriosus, need medical check-ups every 3 to 5 years, either in a non-specialized setting or at shared care facilities (5,6).

When patients with CHD transition from adolescence to adulthood, they should be transferred to the most appropriate adult-focused facility without interruption (10). However, studies in Canada (11,12), Germany (4), the United Kingdom (13), and the United States (14) have demonstrated that 21% to 76% of adolescents with CHD are either lost to follow-up or experience lapses in care after leaving pediatric cardiology. To the best of our knowledge, no studies have comprehensively assessed the settings of care in which adolescents with CHD receive care after leaving pediatric cardiology. Therefore, the aims of the present study were: 1) to determine the transfer destinations of adolescents with CHD after leaving pediatric cardiology; 2) to determine the proportion of patients with no follow-up and no appropriate follow-up after leaving pediatric cardiology; and 3) to explore the determinants of no-follow-up and no appropriate follow-up.

Methods

Setting

As part of the Switch² (Self-Management and Well-being Improvements by Transitioning Adolescents With Chronic Disorders in Hospital and at Home) research program (15), we conducted a descriptive, observational study at the University Hospitals Leuven (Leuven, Belgium). Belgium is a small country with a high population density. Belgium currently has 7 tertiary care centers for pediatric cardiology (16). The pediatric cardiology department of the University Hospitals Leuven cares for 27% of Belgian patients with CHD. At this center, it is standard practice to transfer patients from pediatric cardiology to adult-focused services when they reach 16 years of age, unless they are medically unstable. Because the pediatric CHD and ACHD programs are located in the same building, transferring patients and medical information is easy. Furthermore, both programs share one database of the clinical follow-up of patients. Although transfer from pediatrics to adult-focused care is well established, we do not have a formal transition program that prepares adolescents for the transfer and to take responsibility of their own care (17).

Study population

Eligible patients were adolescents with CHD, which was defined as structural abnormalities of the heart or intrathoracic great vessels that are actually or potentially of functional significance (18); were born between 1984 and 1988; and had one or more cardiac consults in pediatric cardiology between 2000 and 2004. The rationale for selecting these patients is that they had at least 1 outpatient visit at pediatric cardiology during adolescence, showing that they were not considered to be cured in childhood. Because they were 21 years of age or older in 2009, it could be assumed that all patients were cared for in adult-focused care facilities. We excluded patients who had died and

those who had morphologically normal hearts, Wolff-Parkinson-White syndrome or cardiac arrhythmia without structural defects, non-cardiac congenital defects, or pulmonary hypertension without structural anomalies. Heart transplant recipients were also excluded. On the basis of the center's database and outpatient appointment lists, we were able to identify all patients who met the inclusion and exclusion criteria. We included all 813 patients who were eligible for inclusion in this study. Nineteen patients moved abroad and were excluded for statistical analysis because we could not obtain information about their current level of care. Hence, the final sample comprised 794 patients. Socio-demographic and clinical characteristics of this sample are presented in **Table 3.1**.

Definitions

To categorize patients according to their primary heart defect, we used a modified version of the hierarchy of heart defects developed by the CONCOR (CONgenital COR Vitia) project, an initiative to form a national registry of patients with CHD in the Netherlands (18). The modifications are detailed elsewhere (16). In **Table 3.1**, the heart defects are rank ordered according to the CONCOR classification scheme. Furthermore, using the criteria of the Task Force 1 of the 32nd Bethesda Conference, we categorized patients according to the complexity of their heart defects (simple, moderate, and complex) (20).

Transfer destinations were defined based on the three levels of CHD care described by Deanfield et al. (6): specialist care, shared care, and non-specialist care. For the purposes of the present study, we subdivided specialist care into pediatric cardiology care, ACHD care, and care at satellite centers. The latter type of care refers to local hospitals that have a CHD cardiologist-operated outpatient clinic. Shared care is defined as care performed by general cardiologists who see patients with CHD but who send reports to a specialist centers for passing on information and obtaining clinical advice, if needed. Finally, non-specialist care is defined as care by general adult cardiologists who do not send follow-up reports to a specialist centers or care provided by general practitioners.

For inferential statistics, patients were noted as having no follow-up if they indicated that they were currently not in cardiac follow-up or if they could not be contacted by mail or phone. Minimal levels of care were determined according to the type and complexity of heart defects (5,6). The guidelines of Task Force 4 of the 32nd Bethesda Conference (5), with a few exceptions, are applied by our pediatric and ACHD cardiologists. Their expert opinions were used to determine the appropriate minimal level of care for our patients. Patients were noted as being in appropriate follow-up if they received follow-up in a setting that was minimally required or more specialized.

Table 3.1: Sociodemographic and Clinical Characteristics of 794 Patients with CHD

Socio-demographic and clinical characteristics		n (%)
Year of birth	1984	147 (18.5)
	1985	126 (15.9)
	1986	175 (22.0)
	1987	158 (19.9)
	1988	188 (23.7)
Sex	Male	421 (53.0)
	Female	373 (47.0)
Primary CHD diagnosis	Hypoplastic left-heart syndrome	1 (0.1)
	Univentricular physiology	14 (1.8)
	Tetralogy of Fallot	72 (9.1)
	Pulmonary atresia with VSD	0 (0)
	Pulmonary atresia without VSD	2 (0.3)
	DORV	18 (2.3)
	DILV	1 (0.1)
	Truncus arteriosus	3 (0.4)
	TGA	31 (3.9)
	Congenitally-corrected TGA	4 (0.5)
	Coarctation of the aorta	90 (11.3)
	AVSD	48 (6.0)
	ASD type I	10 (1.3)
	Ebstein malformation	3 (0.4)
	Pulmonary valve abnormality	94 (11.8)
	Aortic valve abnormality	106 (13.4)
	Aortic abnormality	14 (1.8)
	Left ventricle outflow tract obstruction	22 (2.8)
	ASD type II	60 (7.6)
	VSD	118 (14.9)
	Mitral valve abnormality	51 (6.4)
	Pulmonary vein abnormality	10 (1.3)
	Other	22 (2.8)
Complexity of primary CHD diagnosis	Simple	272 (34.3)
	Moderate	448 (56.4)
	Complex	74 (9.3)
Prior interventions	No intervention	301 (37.9)
	Only catheter intervention	60 (7.6)
	Only surgical intervention	379 (47.7)
	Both catheter and surgical intervention	54 (6.8)
Distance from home to University Hospitals Leuven (km)	0-49	235 (29.6)
	50-99	407 (51.3)
	100-149	125 (15.7)
	150-199	26 (3.3)
	>200	1 (0.1)

Values are n (%); ASD= atrial septal defect; AVSD= atrioventricular septal defect; CHD= congenital heart disease; DILV= double-inlet left ventricle; DORV= double-outlet right ventricle; TGA= transposition of the great arteries; VSD= ventricular septal defect

Procedure

For 676 patients, data on setting of cardiac follow-up were obtained from our hospital information system. The remaining 137 patients received an information letter, including an informed consent form, requesting information about their current follow-up settings. We telephoned patients as a reminder. Nine patients could not be contacted by mail or telephone; they were untraceable. Sex, year of birth, primary CHD diagnosis, CHD complexity, prior cardiac surgery, prior catheter interventions, and distance from patients' home to the University Hospitals Leuven was determined on the basis of the patients' medical records and additional data.

The institutional review board of the University Hospitals Leuven approved the study protocol. The study was performed in accordance with ethical standards, as described in the 2002 Declaration of Helsinki.

Statistical analysis

Data were analyzed using SPSS version 17.0 (SPSS, Inc., Chicago, Illinois). Nominal and ordinal data are presented as absolute numbers and percents. To determine sociodemographic and clinical variables associated with no follow-up and no appropriate follow-up, we performed multivariable logistic regression analysis using a backward stepwise method. Results are reported as odds ratios (ORs) and 95% confidence intervals (CIs). All tests were 2-sided, and a p value of 0.05 was used as a cut-off for statistical significance.

Results

Transfer destinations

Out of the 794 patients included in the study, a total of 627 adolescents with CHD (79.0%) received follow-up at a tertiary care center. Of these, 613 (97.8%) were transferred to ACHD programs, and 14 (2.2%) were still in follow-up at pediatric cardiology (**Figure 3.1**). In addition, 57 patients (7.2%) received follow-up at a satellite center. Hence, altogether, 86.1% of the patients continued to receive specialist care when they reached young adulthood. Fifty-two adolescents (6.5%) received cardiac follow-up from a general adult cardiologist. In 29 of the 52 cases (55.8%), cardiologists sent exam reports to CHD specialists; this type of follow-up was considered to be shared care.

Forty-nine patients (6.2%) had no cardiac follow-up after leaving pediatric cardiology, and 9 patients (1.1%) were untraceable. Hence, 58 of the 794 patients included (7.3%) were considered to be lost to follow-up. More specifically, 2 of the 74 patients with complex (2.7%), 31 of the 448

patients with moderate (6.9%), and 25 of the 272 patients with simple (9.2%) heart defects were no longer in cardiac follow-up.

Minimal level of cardiac care

On the basis of primary diagnosis and complexity of the congenital heart lesions, we subdivided patients into 3 groups relating to the minimal level of care they should receive. The first group consisted of patients who should receive specialist care exclusively (**Table 3.2**). This level of care can be performed at pediatric cardiology, ACHD programs, or satellite centers. In all, 225 patients (90%) received specialist care. Ten patients (4%) received shared care, 9 patients (3.6%) received non-specialist care, 5 patients (2%) were no longer in follow-up, and 1 (0.4%) patient was untraceable. These latter levels of care were suboptimal for these patients.

Among 247 patients who should receive specialist or shared care, 215 patients (87%) received specialist care and 7 patients (2.8%) received shared care (**Table 3.3**). Four patients (1.6%) received nonspecialist care, 17 patients (6.9%) were not in follow-up, and 4 patients (1.6%) were untraceable.

Of the 297 patients for whom nonspecialist care would be sufficient, 244 patients (82.2%) continued specialized follow-up, 12 patients (4%) received shared care, and 10 patients (3.4%) received nonspecialist care. Twenty-seven patients (9.1%) were not in cardiac follow-up, and 4 patients (1.3%) were untraceable (**Table 3.4**).

Altogether, our data revealed that 81 patients (10.2%) did not receive follow-up at the recommended level of care. These patients were considered as receiving no appropriate cardiac follow-up.

Figure 3.1: Transfer destinations in 794 patients with CHD

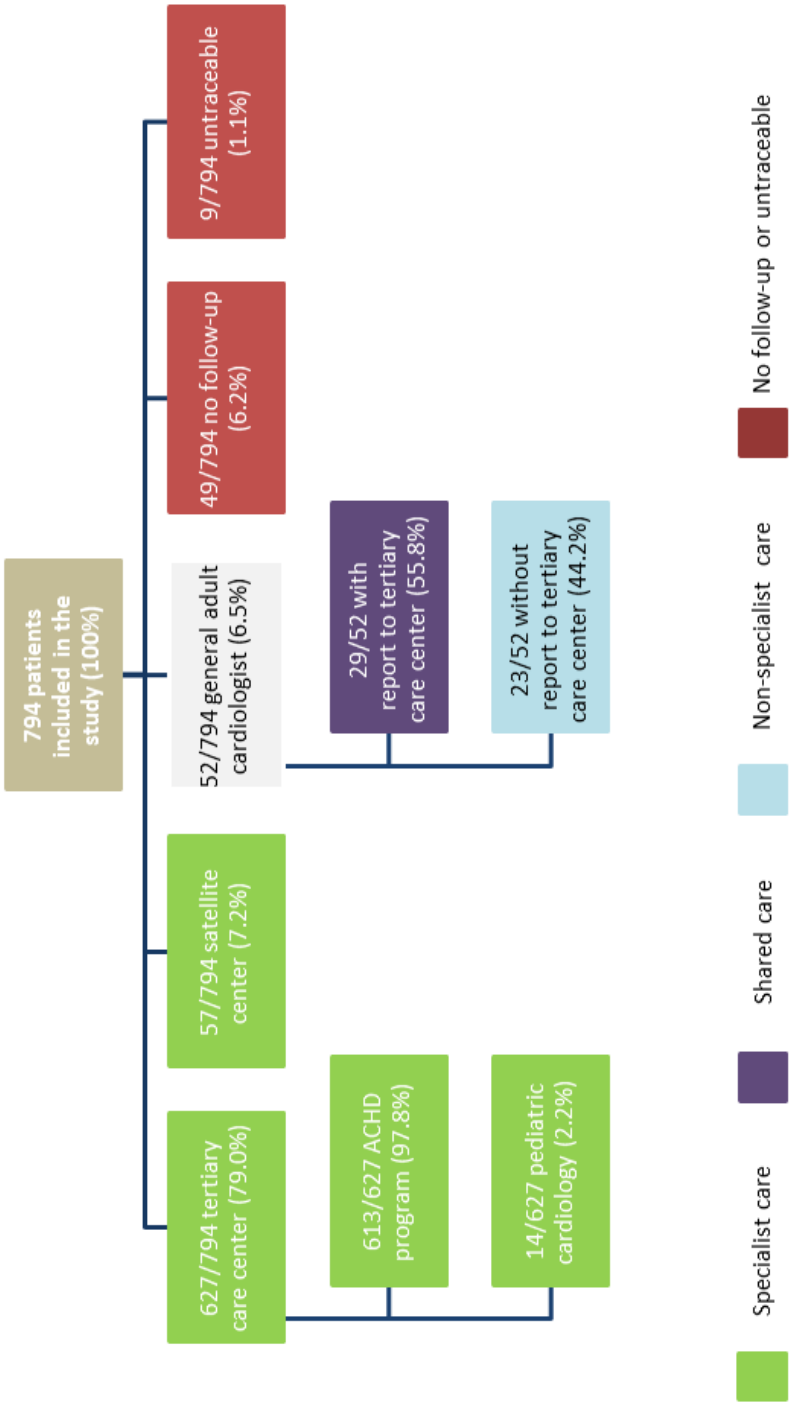

Table 3.2: Congenital Heart Defects Requiring Specialist Care

Type of CHD	Specialist care	Shared care	Non-specialist care	No follow-up	Not traceable
Hypoplastic left-heart syndrome	1				
Univentricular heart	14				
Pulmonary atresia without VSD	2				
DORV	16		2*		
DILV	1				
Truncus arteriosus	3				
TGA	28		2*		1*
Congenitally corrected TGA	3			1*	
Coarctation of the aorta	80	5*	2*	3*	
Aortic abnormality	11	2*		1*	
Tetralogy of Fallot	66	3*	3*		
Proportion of patients in each setting of follow-up	90.0% (n=225)	4.0% (n=10)*	3.6% (n=9)*	2.0% (n=5)*	0.4% (n=1)*

** Patients who did not receive the minimal level of cardiac care. Abbreviations as in Table 3.1.*

Table 3.3 Congenital Heart Defects Requiring Specialist or Shared Care

Type of CHD	Specialist care	Shared care	Non-specialist care	No follow-up	Not traceable
AVSD	45		1*	2*	
ASD type I	7	1	1*		1*
Ebstein malformation	3				
Associated pulmonary valve abnormality	31	1	1*	3*	1*
Left ventricle outflow tract obstruction	19			3*	
VSD	29	2		3*	2*
Associated mitral valve abnormality	9			2*	
Pulmonary vein abnormality	10				
Isolated pulmonary valve abnormality	50	3	1*	3*	
Other	12			1*	
Proportion of patients in each setting of follow-up	87,0% (n=215)	2,8% (n=7)	1.6% (n=4)*	6,9% (n=17)*	1,6% (n=4)*

** Patients who did not receive the minimal level of cardiac care. Abbreviations as in Table 3.1.*

Table 3.4: Congenital Heart Defects for Which Non-specialist Care Is Sufficient

Type of CHD	Specialist care	Shared care	Non-specialist care	No follow-up	Not traceable
Isolated aortic valve abnormality	47	2	4	2*	
Associated ASD type II	27	1	1	2*	
Isolated ASD type II	27	1		1*	
VSD repaired without residua	72	2	2	4*	2*
Isolated mitral valve abnormality	25	2	2	9*	2*
Associated aortic valve abnormality	40	3	1	7*	
Other	6	1		2*	
Proportion of patients in each setting of follow-up	82.2% (n=244)	4.0% (n=12)	3.4% (n=10)	9.1% (n=27)*	1.3% (n=4)*

** Patients who did not receive the minimal level of cardiac care. Abbreviations as in Table 3.1.*

Factors associated with 'no follow-up' and 'no appropriate follow-up'

We investigated the association between five socio-demographic and clinical variables and no follow-up and no appropriate follow-up: sex, prior heart surgery, prior catheter intervention, complexity of heart defect, and distance from patients' homes to our hospital. Independent correlates of no cardiac follow-up after leaving pediatric cardiology were male sex (OR:1.80; 95%CI:1.02 to 3.17) and no prior heart surgery (OR: 5.97; CI: 3.04 to11.72). No appropriate level of cardiac follow-up after leaving pediatric cardiology was associated with male sex (OR: 1.63; CI: 1 to 2.63), no prior heart surgery (OR: 3.30; CI: 1.88 to 5.77,); and greater complexity of CHD (OR: 1.61; CI: 1.04 to 2.49).

Discussion

Continuing follow-up is important for many patients with CHD. However, studies have shown that numerous patients are lost to follow-up or have lapses in care after leaving pediatric cardiology (4,11-14). Furthermore, not all patients receive the optimal level of care. Therefore, we investigated the destinations of transfer in adolescents with CHD, determined the proportion of patients with no follow-up and no appropriate follow-up after they leave pediatric cardiology, and sought correlates of no-follow-up and no appropriate follow-up.

Comparing our findings with published data, the situation at our center appears to be substantially better than in other Western countries. To date, five studies have described the proportion of patients lost to follow-up and/or experiencing lapses in care after leaving pediatric cardiology (21). Reid et al. (11) investigated medical care in 360 patients aged 19-to 21 years with complex CHD. These patients were followed up in pediatric cardiology at the Hospital for Sick Children in Toronto, Ontario, Canada, before the age of 18 years. The investigators defined successful transfer as patients attending ≥1 appointment at a Canadian ACHD center. In this study, 53% of the patients did not successfully transfer, and >25% had no cardiac appointments after the age of 18 years.

At the German Heart Center in Munich, Germany, Wacker et al. (4) evaluated the rate and outcomes of adults with CHD lost to follow-up at their institution. Patients were selected from the CHD program registry (n>10,500). This population included a broad spectrum of CHD. Loss to follow-up was defined as patients' failing to return for follow-up visits to their center for >5 years. The investigators found that >76% of patients were lost to follow-up.

In another study, de Bono and Freeman (13) assessed 59 patients with coarctation of the aorta. This study was performed in the United Kingdom at a local ACHD clinic without on-site

cardiothoracic surgery or pediatric cardiology facilities. Patients who were in follow-up at the ACHD clinic at the time of the study, but who were not being seen at other cardiac clinics for a period of ≥2 years, were considered lost to follow-up. Thirty-nine percent of the patients had ≥1 episode of lost to follow-up.

Yeung et al. (14) conducted a study in Denver, Colorado, that determined the proportion of patients (with a moderate or complex heart defects) who experienced lapses in medical care after leaving pediatric cardiology. A lapse in care was defined as a >2-year interval between leaving pediatric cardiology and presentation at the ACHD clinic. In 63% of patients, lapses of care were observed, with a median interval duration of 10 years.

Mackie et al. (12) conducted a population-based investigation in Quebec, Canada, of 643 patients diagnosed with CHD before 6 years of age and currently 22 years old. Lack of follow-up, defined as the absence of an outpatient assessment by a cardiologist, was retrieved from the physician billing database. This study revealed that 61% of the patients failed to receive cardiac follow-up after their 18th birthdays. Subgroup analyses showed that 47% of patients with moderate or complex heart defects (mild defects were excluded) were lost to follow-up after their 18th birthdays, whereas 21% of patients with complex lesions (mild and moderate defects were excluded) were lost to follow-up.

Generally, a wide variation in percents of patients lost to follow-up or those with lapses of care has been observed. However, these data are not comparable, because the studies differed substantially in terms of definition of loss to follow-up, study population, inclusion criteria, recruitment setting (pediatric cardiology, ACHD clinic, or population based), data collection methods (database or retrospective evaluation), and follow-up period. This likely resulted in an underestimation and overestimation of the proportions of patients lost to follow-up. For example, de Bono and Freeman (13) and Yeung et al. (14) recruited patients at ACHD clinics. By doing so, they underestimated the problem of loss to follow-up, because patients not under medical surveillance were excluded in their studies. In contrast, Wacker et al. (4) included all patients recorded in their center's registry and considered patients to be lost to follow-up if they did not have checkups in that specific center. Consequently, they probably overestimated lost to follow-up, because a substantial number of patients with mild defects may not have needed ongoing cardiac follow-up, and some patients received cardiac follow-up at other centers.

Regardless of the limited comparability across these studies, our study demonstrated a considerably lower proportion of loss to follow-up. There are several explanations. First, at our center, pediatric cardiology and an ACHD program are located in the same building. Hence, patients

do not have to go to another hospital when being transferred to adult care. Second, pediatric and ACHD cardiologists at our center use the same medical records, hospital information system, and database. This facilitates transfer of medical information. Third, to keep patients under medical surveillance, our clinic sends outpatient visit reminders to the patients, according to the proposed frequency of follow-up visits. Non-responding patients will receive up to 3 reminders. Fourth, Belgium has a compulsory health insurance system, covering almost the entire population. Therefore, noninsurance or underinsurance is no barrier for patients wanting to obtain the care needed. Fifth, there is no mandatory general practitioner gate-keeping system in Belgium, resulting in easy access to tertiary care, which increases the accessibility of CHD care. Finally, Belgium is a small country with a high population density. Hence, the distances from patients' homes to specialized centers are relatively short. In our sample, 80% of the patients lived <100 km (<62 mi) from our hospital.

To what extent these factors affected our findings is unknown. However, obviously not only patient-related factors but also healthcare system- related and hospital-related factors have an impact on successful continuation of cardiac follow-up when patients reach adulthood. Indeed, the availability and structure of CHD programs will have an impact on how care is provided (22,23). To address this issue, we are currently preparing the INTERCHANGE (INTERnational study on the Continuation of Heart health checks in young Adults with coNGEnital heart disease) study, an international study on healthcare-related, hospital-related, and patient-related determinants of lack of cardiac follow-up in adulthood. This will be an observational study using a multilevel approach, with data collection at three levels: country, center, and patient levels. Across Europe and North America >20 centers will participate, including about 7,500 patients.

In the present study, we focused on the minimal level of care. We found that 10.2% of our patients did not receive follow-up at the minimally recommended level. However, we also observed that the level of care exceeded the guidelines in many patients (5). For instance, in the group of patients for whom nonspecialist care is sufficient (**Table 3.4**), 86.3% of patients received specialist or shared care. If the ACHD program is saturating, there would be an opportunity to discharge patients with mild heart defects to lower levels of care. So far, we have not done so, because our pediatric cardiology and ACHD programs are located in a teaching hospital. In terms of training of cardiology fellows, it is considered to be appropriate to have exposure to the entire spectrum of CHD.

Study limitations

First, this study was conducted in one tertiary center with a specific structure and located in a particular healthcare system. Thus, our results are not generalizable. Second, this study mainly applied the Task Force 4 recommendations for cardiac follow-up (5). These recommendations are not completely consistent with, for instance, European guidelines (6). Application of European guidelines would likely result in different findings. Third, only patient-related correlates of no follow-up and no appropriate follow-up were investigated, leaving healthcare system-related and hospital-related factors unaddressed. The planned INTERCHANGE study, however, will address these factors.

Conclusion

Only 7.3% of our patients with CHD were no longer in cardiac follow-up after leaving pediatric cardiology. Of the patients with complex, moderate and simple CHD, 2.7%, 6.9%, and 9.2%, respectively, were no longer in follow-up. According to international guidelines, 10.2% of our patients did not receive follow-up at the minimally recommended level. No follow-up was associated with male sex and no prior heart surgery. No appropriate follow-up was correlated with male sex, no prior heart surgery, and greater complexity of CHD. Our results are substantially better than those in other Western countries. Firm explanations for the observed differences will be determined in our future study on healthcare-, hospital- and patient-related determinants.

References

1. Hoffman JI, Kaplan S. The incidence of congenital heart disease. J Am Coll Cardiol 2002;39:1890-1900.
2. Moons P, Bovijn L, Budts W, Belmans A, Gewillig M. Temporal trends in survival into adulthood among patients born with congenital heart disease from 1970-1992 in Belgium. Circulation, 2010;22:2264-2272.
3. Knauth A, Verstappen A, Reiss J, Webb GD. Transition and transfer from pediatric to adult care of the young adult with complex congenital heart disease. Cardiol Clin 2006;24:619-629.
4. Wacker A, Kaemmerer H, Hollweck R et al. Outcome of operated and unoperated adults with congenital cardiac disease lost to follow-up for more than five years. Am J Cardiol 2005;95:776-779.
5. Landzberg MJ, Murphy DJ Jr, Davidson WR Jr. et al. Task force 4: organization of delivery systems for adults with congenital heart disease. J Am Coll Cardiol 2001;37:1187-1193.
6. Deanfield J, Thaulow E, Warnes C et al. Management of grown up congenital heart disease. Eur Heart J 2003;24:1035-1084.
7. Foster E, Graham TP Jr, Driscoll DJ et al. Task force 2: special health care needs of adults with congenital heart disease. J Am Coll Cardiol 2001;37:1176-1183.
8. Murphy DJ Jr., Foster E. ACCF/AHA/AAP recommendations for training in pediatric cardiology. Task force 6: training in transition of adolescent care and care of the adult with congenital heart disease. J Am Coll Cardiol 2005;46:1399-1401.
9. Warnes CA, Williams RG, Bashore TM et al. ACC/AHA 2008 Guidelines for the Management of Adults with Congenital Heart Disease: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (writing committee to develop guidelines on the management of adults with congenital heart disease). J Am Coll Cardiol 2008;52:e143-e263.
10. Hilderson D, Saidi AS, Van Deyk K et al. Attitude toward and current practice of transfer and transition of adolescents with congenital heart disease in the United States of America and Europe. Pediatr Cardiol 2009;30:786-793.
11. Reid GJ, Irvine MJ, McCrindle BW et al. Prevalence and correlates of successful transfer from pediatric to adult health care among a cohort of young adults with complex congenital heart defects. Pediatrics 2004;113:e197-e205.
12. Mackie AS, Ionescu-Ittu R, Therrien J, Pilote L, Abrahamowicz M, Marelli AJ. Children and adults with congenital heart disease lost to follow-up: who and when? Circulation 2009;120:302-309.
13. de Bono J, Freeman LJ. Aortic coarctation repair-lost and found: the role of local long term specialised care. Int J Cardiol 2005;104:176-183.
14. Yeung E, Kay J, Roosevelt GE, Brandon M, Yetman AT. Lapse of care as a predictor for morbidity in adults with congenital heart disease. Int J Cardiol 2008;125:62-65.
15. Katholieke Universiteit Leuven. The SWITCH^2 research program. Available at: http://www.kuleuven.be/switch2/. Accessed July 22,2010.
16. Moons P, Sluysmans T, De Wolf D et al. Congenital heart disease in 111 225 births in Belgium: birth prevalence, treatment and survival in the 21st century. Acta Paediatr 2009;98:472-477.
17. Moons P, Pinxten S, Dedroog D et al. Expectations and experiences of adolescents with congenital heart disease on being transferred from pediatric cardiology to an adult congenital heart disease program. J Adolesc Health 2009;44:316-322.
18. Mitchell SC, Korones SB, Berendes HW. Congenital heart disease in 56,109 births. Incidence and natural history. Circulation 1971;43:323-332.
19. Vander Velde ET, Vriend JW, Mannens MM, Uiterwaal CS, Brand R, Mulder BJ. CONCOR, an initiative towards a national registry and DNA-bank of patients with congenital heart disease in the Netherlands: rationale, design, and first results. Eur J Epidemiol 2005;20:549-557.
20. Warnes CA, Liberthson R, Danielson GK et al. Task force 1: the changing profile of congenital heart disease in adult life. J Am Coll Cardiol 2001;37:1170-1175.

21. Moons P, Hilderson D, Van Deyk K. Implementation of transition programs can prevent another lost generation of patients with congenital heart disease. Eur J Cardiovasc Nurs 2008;7:259-263.
22. Moons P, Engelfriet P, Kaemmerer H, Meijboom FJ, Oechslin E, Mulder BJ. Delivery of care for adult patients with congenital heart disease in Europe: results from the Euro Heart Survey. Eur Heart J 2006;27:1324-1330.
23. Moons P, Meijboom FJ, Baumgartner H, Trindade PT, Huyghe E, Kaemmerer H. Structure and activities of adult congenital heart disease programmes in Europe. Eur Heart J 2010;31:1305-1310.

4 IMPLEMENTATION OF GUIDELINES FOR THE MANAGEMENT OF ACHD FOLLOW-UP CARE

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Abstract

Objective: Although different guidelines on ACHD care advocate for lifetime cardiac follow-up, a critical appraisal of the guideline implementation is lacking. We investigated the implementation of the ACC/AHA 2008 guidelines for Adult Congenital Heart Disease (ACHD) follow-up by investigating the type of healthcare professional; care setting and frequency of outpatient visits in young adults with CHD. Furthermore, correlates for care in line with the recommendations or untraceability were investigated.

Methods: A cross-sectional, monocentric, observational study was conducted, including 306 patients with CHD who had a documented outpatient visit at pediatric cardiology before age 18y.

Results: Two-hundred ten patients (68.6%) were in cardiac follow-up; 20 (6.5%) withdrew from follow-up and 76 (24.9%) were untraceable. Overall, 198 patients were followed-up in tertiary care, one quarter (n=52) of which were seen at a formalized ACHD care program, and three quarters (n=146) remained at pediatric cardiology. Of those followed in formalized ACHD and pediatric cardiology care, the recommended frequency was implemented in 94.2% and 89%, respectively (p=.412). No predictors for the implementation of the guidelines were identified. Risk factors for becoming untraceable were: none or lower number of heart surgeries; health insurance issues; and non-white ethnicity.

Conclusions: A significant number of adults continue to be cared for by pediatric cardiologists, indicating that transfer to adult-oriented care was not standard practice. Frequency of follow-up for the majority of patients was in line with the ACC/AHA 2008 guidelines. A considerable proportion of young adults were untraceable in the system, which makes them vulnerable for discontinuation of care.

Introduction

The number of adults born with congenital heart disease (CHD) is expanding substantially (1). Longitudinal studies have demonstrated that although mortality rates have declined spectacularly over the past decades, morbidity rates in this population tend to be high. Since most adults with CHD face a number of potential complications and reoperations as they age, lifetime cardiac follow-up is required in most patients (2).

In childhood, patients are standardly followed-up in pediatric cardiology programs. As patients grow into adulthood, changing healthcare needs advocate a transfer from pediatric to adult healthcare services (3). Several professional organizations for both pediatric and adult care propose educational programming and ultimately a transfer of care to provide continuous care in accordance with the developmental stage of patients diagnosed with CHD (3;4). Furthermore, a recent population-based study was the first to demonstrate a beneficial effect of increased referral rates to ACHD care centers on mortality in adults with CHD (5).

The requirements for adult CHD (ACHD) follow-up care have been outlined by several cardiovascular task forces. These documents detailed the type of professionals, the preferred setting and the frequency at which surveillance should be provided (6). The ACC/AHA 2008 management guidelines supported the framework for the organization of ACHD care that was developed at the 32nd Bethesda conference in 2001 (7;8). A hierarchical algorithm for ACHD care provision is based primarily upon the anatomical classification of the diagnosed heart lesion(s). Patients diagnosed with a simple defect are generally recommended to receive follow-up care in the general medical community every 3 to 5 years. This level of non-specialized care can be provided by a primary care physician or a community cardiologist. A more intensive frequency of visits is advised for patients with moderately to highly complex defects, as they have an increased vulnerability to develop comorbidities. In case of moderately complex defects, collaborative care is preferably provided in specialist centers, even though shared care between community providers and ACHD specialists can be recommended if the course of the condition is uncomplicated. Care exclusively provided by cardiologists who are specialized in ACHD is proposed every 6-12 months for patients with highly complex CHD (8). Hence, these recommendations are in line with the stratified model of ACHD care delivery, as proposed by the European Society of Cardiology (6).

Despite the well-established importance of continuous care provision for ACHD patients, previous studies have demonstrated that 7 to 76% of patients experienced care gaps or were lost to cardiac follow-up during adulthood (9-16). These studies have shown that the provision of

continuous care to young people with CHD remains a largely unfulfilled need in Western countries. The implications can, however, be far-reaching as demonstrated by the threefold likelihood of needing urgent cardiac interventions in patients experiencing a care gap (9).

Although guidelines on the provision of ACHD follow-up care were published about a decade ago, implementation of these recommendations in clinical practice has yet to be examined. Therefore, this cross-sectional study describes the implementation of ACC/AHA 2008 guidelines for ACHD care through the investigation of the type of professional, the setting and the frequency at which outpatient visits are performed in young adults with CHD. Furthermore, this paper explores the profile of patients who received care in line with the recommendations and compares this with the profile of patients who appeared untraceable in the healthcare system.

Material and methods

Setting

This observational study was conducted at a large free-standing pediatric hospital (Boston Children's Hospital) in the United States of America that has an outpatient cardiology volume of > 23,000 visits per year. There is an ACHD care clinic onsite and a transitioning liaison helping patients navigate from pediatric cardiology to ACHD care. Subjects for study inclusion were identified by searching institutional and departmental databases.

Study population

Eligible patients were selected from the pediatric cardiology outpatient clinic list overviewing all visits performed between 2001-2005. Based on the following criteria, patients were selected for participation: a confirmed diagnosis of CHD, defined as "*a gross structural abnormality of the heart and/or the intrathoracic great vessels that is actually or potentially of functional significance*" (17); born in 1987 (aged 23y in 2010); at least one documented outpatient pediatric cardiology visit at the institution between 2001-2005 (before age 18y); and living in the region of New England (USA) at the time of data collection.

Data collection

A clinical research form was completed after reviewing patient's medical files. Information on sex, highest level of education, primary CHD diagnosis, and type of health insurance was collected based on chart review at the last outpatient visit. The ethnicity of patients was determined based on self-report. Based on the ZIP code of the place of residence, the travel distance (miles) to the nearest

ACHD center was calculated. Furthermore, the number of cardiac interventions performed in the past was determined. Interventions of cardiac ablation and implantation of a pacemaker or a cardioverter defibrillator were included, whereas intracardiac electrophysiology studies were not taken into account.

Furthermore, a four phase-approach was used to determine the setting and frequency of the outpatient visits. During phase one, the hospital's cardiology database was checked to determine if patients were currently in follow-up at the institution. Subsequently, the Social Security Death Index was checked to exclude deceased patients. If data were missing or unavailable from chart review, and the mortality status remained undetermined, the last known cardiologist was contacted. Finally, if no additional information on follow-up could be provided by this cardiologist, the patient was contacted through mail or telephone using the patient's last known contact details.

This study was approved by the Boston Children's Hospital Center for Clinical Investigation and was performed in accordance with the 2002 Declaration of Helsinki.

Definitions

The CHD diagnosis was determined based on chart review. The primary heart lesion was rank ordered using a modified CONCOR (CONgenital COR Vitia) classification (18). The defect was anatomically defined as simple, moderate or complex using Task Force 1 (7). Patients were considered to be currently in follow-up if an outpatient visit could be documented using the aforementioned four-phase protocol. Furthermore, patients were categorized as not being in cardiac follow-up if a complete cessation of cardiac care was confirmed. If at stage four, the patient could not be contacted, then this patient was considered to be untraceable and no further information on the cardiac follow-up care was collected.

For the group of patients who were currently in follow-up, detailed information on the setting and frequency of scheduled visits was derived. The setting of care was subdivided into four groups: care provided by (a) a pediatric cardiologist exclusively, (b) an ACHD cardiologist exclusively, (c) a general community cardiologist in collaboration with a specialized CHD team (shared care), or (d) a general community cardiologist solely.

To assess the implementation of the ACC/AHA 2008 guidelines, the setting and frequency of visits was compared to the recommended setting of care and frequency of visits (19). Care was considered to be not in line with the current ACC/AHA 2008 recommendations, if the setting or frequency of cardiac follow-up visits was not in accordance to the minimally recommended level or frequency.

Statistical analysis

Data were analyzed using SPSS 20.0 (SPSS, Inc., Chicago, Illinois). Nominal and categorical variables were presented using absolute numbers and proportions. Median values were reported for the non-normally distributed continuous variables. To explore the profile of patients receiving care at the recommended setting and frequency and patients found to be untraceable, two multivariable logistic regression models were analyzed (forced entry method). Assumptions of linearity, multicollinearity and independence of errors were checked and found to be met. Results were reported as odds ratios (ORs) and 95% confidence intervals (CIs). All statistical tests were 2-sided and a p-value of 0.05 was used as a cut-off point for statistical significance.

Results

Sample characteristics

A total of 327 patients were eligible. However, one patient explicitly opted-out of further inquiries. Twenty patients moved out of New England, leaving 306 patients (94%) included in data analysis. Demographic and clinical characteristics are provided in **Table 4.1**.

Table 4.1: Demographic and clinical sample characteristics (n=306)

Variables	n (%)
Sex	
male	180 (58.8)
female	126 (41.2)
Primary diagnosis of CHD	
Hypoplastic left-heart syndrome	2 (0.7)
Univentricular physiology	13 (4.2)
Tricuspid atresia	2 (0.7)
Tetralogy of Fallot	29 (9.5)
Pulmonary atresia without VSD	3 (1)
DORV	6 (2)
TGA	28 (9.2)
Congenitally-corrected TGA	2 (0.7)
Coarctation of the aorta	24 (7.8)
AVSD	8 (2.6)
ASD type I	1 (0.3)
Ebstein malformation	3 (1)
Pulmonary valve abnormality	26 (8.5)
Aortic valve abnormality	60 (19.6)
ASD type II	43 (14.1)
VSD	39 (12.7)
Mitral valve abnormality	3 (1.0)
Pulmonary vein abnormality	5 (1.6)
Other diagnoses	9 (2.9)

Morphologic complexity of CHD *(Warnes et al., 2001)*	
Simple	141 (46.1)
Moderate	98 (32)
Complex	67 (21.9)
Prior heart surgery	
No, none	139 (45.4)
Yes, ≥1 surgery performed	167 (54.6)
Median number of surgeries	1
Range	0-7
Prior cardiac catheter-based interventions	
No, none	173 (56.5)
Yes, ≥ 1 catheterization performed	133 (43.5)
Median number of catheter-based interventions	0
Range	0-31
Race/ethnicity	
Caucasian	226 (73.9)
Hispanic/Latino	15 (4.9)
African American	14 (4.6)
Asian	7 (2.3)
Other	44 (14.4)
Highest level of education	
Grades K8-12	9 (2.9)
Graduated high school	101 (33)
Associate's degree	18 (5.9)
Bachelor's degree	108 (35.3)
Special education program	28 (9.2)
Other	4 (1.3)
Unknown (e.g., patient was untraceable)	38 (12.4)

Medical coverage	
Private insurance	169 (55.2)
State aid	53 (17.3)
Self-pay/no insurance	5 (1.6)
Unkown (e.g., patient was untraceable)	79 (25.8)
Travel distance to nearest ACHD center (miles)	
Median	29.9
Range	1-421

ASD = atrial septal defect; AVSD = atrioventricular septal defect; CHD=congenital heart disease; DILV = double-inlet left ventricle; DORV = double-outlet right ventricle; TGA = transposition of the great arteries; VSD = ventricular septal defect

Setting of care for cardiac follow-up

A total of 210 patients (68.6%) were in cardiac follow-up at the age of 23y. However, 20 patients (6.5%) withdrew from follow-up. For 76 patients (24.9%) information on the setting or the frequency of follow-up could not be retrieved from chart review or from last known medial provider. In addition, direct contact with patient via last known phone or address was not possible. Since this latter group of patients could not be contacted, they were considered to be untraceable. Within the group of patients who were in follow-up, the majority (n=198, 64.7%) received cardiac care at tertiary care, while 12 patients (3.9%) were in follow-up with a community cardiologist who had no expertise in CHD care. Although 52 patients (17%) followed in tertiary care were seen at an ACHD program, the majority (n=146, 47.7%) received care exclusively provided by a pediatric cardiologist. Detailed information on the setting of follow-up can be found in **Figure 4.1**.

Implementation of the guideline-recommended setting and frequency of cardiac follow-up

Within the group of patients who were currently in follow-up, only 55 patients (26.2%) were being seen within a formalized ACHD setting of care as proposed by the ACC/AHA 2008 recommendations (**Figure 4.2**). The majority of patients currently in follow-up (89.5%) were being seen at the minimally recommended frequency. Of those followed in formalized ACHD care, the minimally recommended frequency was implemented in 94.2%. For those in pediatric cardiology, this proportion was 89% (p=.412). Taken together, 52 patients (24.8%) of our sample were receiving

follow-up care at both a formalized ACHD care setting and at the frequency as proposed by the guidelines based on the anatomical classification of their primary defect (**Figure 4.2**).

Figure 4.1 Flowchart of Setting of Care for Cardiac Follow-up (n=306)

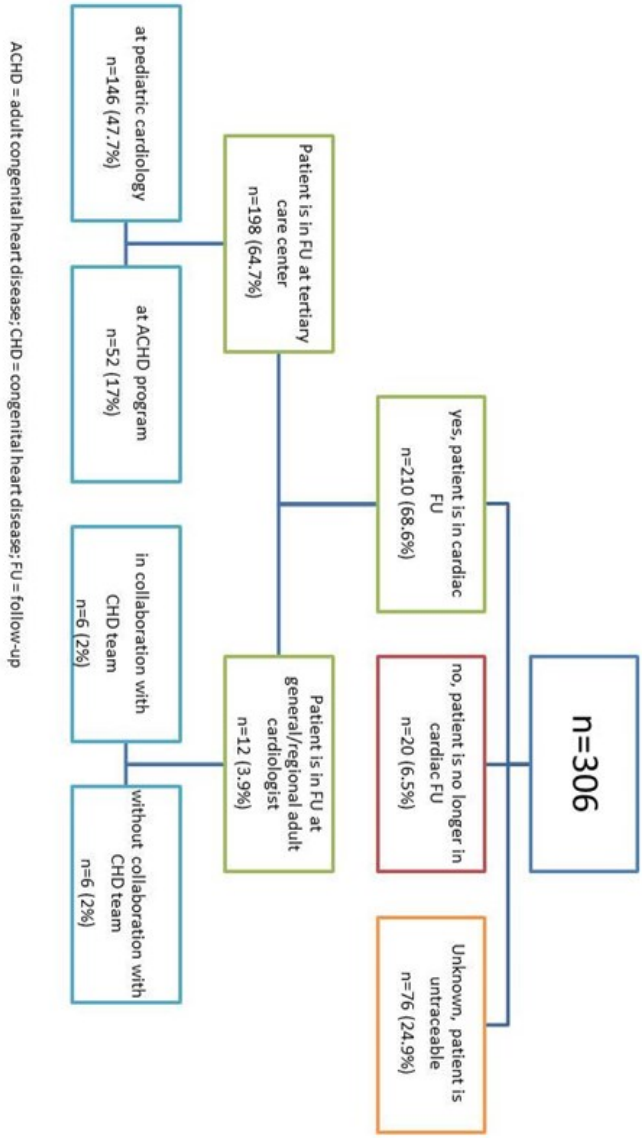
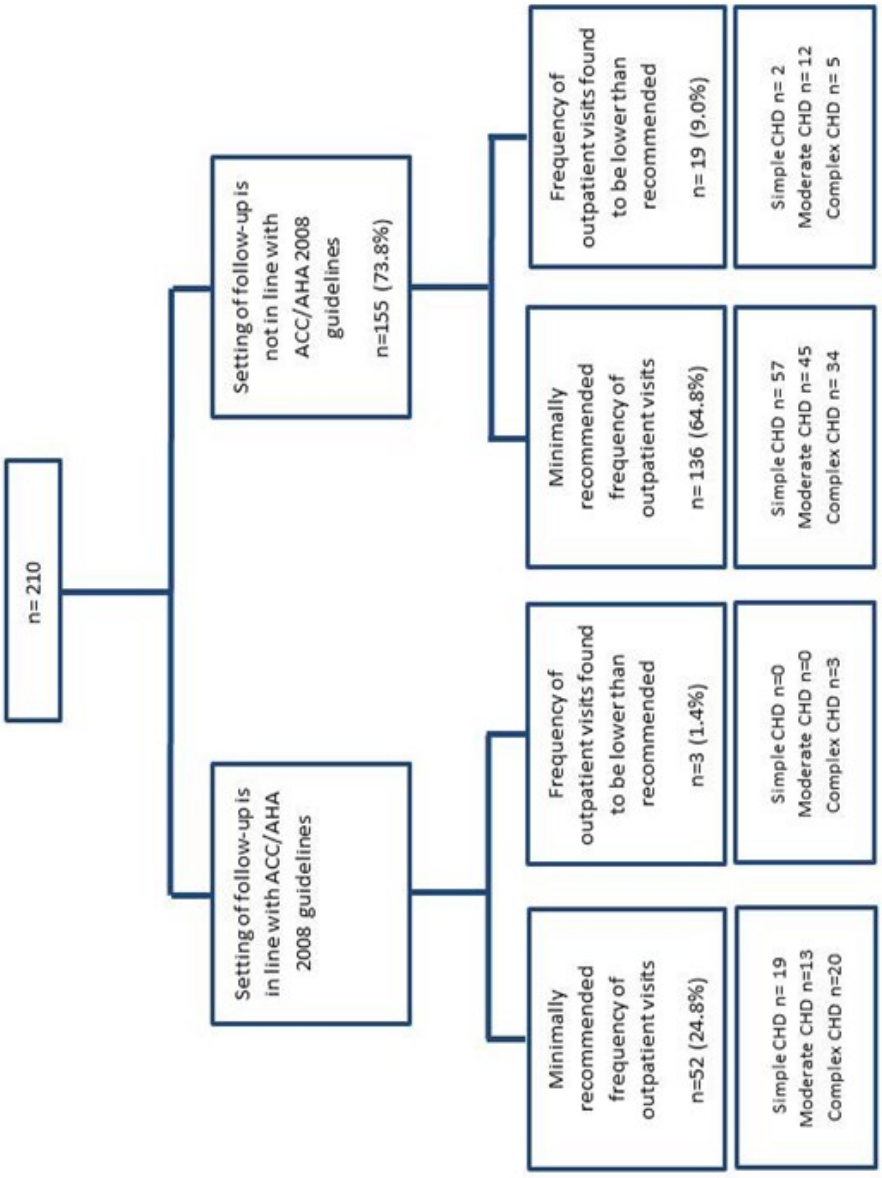


Figure 4.2 Implementation of the Recommended Setting and Frequency of Follow-up (n=210)



Profile of patients receiving follow-up care at the proposed setting and frequency

In this latter group of 52 patients, 19 patients had a simple lesion, 13 were diagnosed with a moderate defect and 20 had a complex defect. Using multivariable logistic regression analysis, the profile of patients who received care within the proposed setting and at the recommended frequency was explored. None of the demographic/clinical variables were, however, found to be significant predictors for receiving such care (**Table 4.2**).

Profile of patients who are untraceable

Patients who are untraceable require our attention, because they may be vulnerable for care gaps. In the group of patients who were untraceable, 65% had a simple defect, 29% were diagnosed with a moderate defect and 7% had complex CHD. Adults who underwent none or a lower number of surgeries in the past (OR=0.4; 95%CI: 0.2-0.7), had no insurance or relied on self-pay (OR=0.1; 95%CI: 0.05-0.2), and were of a non-white ethnicity (OR=3.0; 95% CI: 1.5-6.1) had a significantly increased likelihood to become untraceable (**Table 4.3**). However, sex, anatomical CHD classification, travel distance to the nearest ACHD center and the number of prior interventions, were not found to be predictive of being untraceable.

Table 4.2 Multivariable logistic regression model predicting implementation of the recommended setting and frequency of follow-up visits in adults with CHD (n=210)

Predictor variables	B	S.E.	Exp(B)	95% CI	p-value
Constant	-1.359	0.646	0.257		
Sex	0.185	0.339	1.203	0.619-2.337	0.586
CHD complexity					
simple (reference)	-	-	-	-	0.247
moderate	-0.479	0.429	0.620	0.267-1.436	0.264
complex	0.275	0.517	1.316	0.478-3.627	0.595
Travel distance to nearest ACHD center	-0.005	0.004	0.995	0.987-1.004	0.266
Number of heart surgeries performed in the past	0.020	0.142	1.020	0.773-1.346	0.890
Number of catheter-based interventions performed in the past	0.015	0.052	1.015	0.917-1.123	0.773
Medical insurance*	0.431	0.604	1.538	0.471-5.025	0.476
Ethnicity†	-0.070	0.435	0.932	0.398-2.185	0.872

For the entire cohort, n=210

** 0 indicates no medical insurance or relying on self-pay; 1 indicates patient had medical coverage either through private insurance or state aid*

† 0 indicates patient has white ethnicity; 1 indicates patient has a non-white ethnicity

Table 4.3: Multivariable Logistic Regression Model Predicting Untraceability in Adults with CHD (n=306)

Predictor variables	B	S.E.	Exp(B)	95% CI	p-value
Constant	0.849	0.408	2.337		
Sex	-0.137	0.344	0.872	0.444-1.713	0.692
CHD complexity					
simple (reference)	-	-	-	-	0.446
moderate	0.516	0.417	1.675	0.739-3.795	0.217
complex	0.547	0.764	1.727	0.387-7.719	0.474
Travel distance to nearest ACHD center	-0.009	0.005	0.991	0.982-1.001	0.093
Number of heart surgeries performed in the past	-0.924	0.307	0.397	0.218-0.724	0.003
Number of catheter-based interventions performed in the past	-0.256	0.205	0.774	0.518-1.157	0.212
Medical insurance*	-2.364	0.366	0.094	0.046-0.192	<0.001
Ethnicity†	1.093	0.361	2.983	1.470-6.503	0.002

For the entire cohort, n=306; Nagelkerke R square = 47.3%

** 0 indicates no medical insurance or relying on self-pay; 1 indicates patient had medical coverage either through private insurance or state aid*

† 0 indicates patient has white ethnicity; 1 indicates patient has a non-white ethnicity

Discussion

To the best of our knowledge, this was the first study appraising the implementation of the ACC/AHA 2008 guidelines for ACHD care, several years after their publication. For the purpose of this study, three components of follow-up care were examined: the type of physician providing follow-up, the setting and the respective frequency of visits. We found that 65% of the 23-y old patients were in follow-up at a tertiary center, one-quarter of which was followed-up in the ACHD care program, whereas the remaining three-quarters remained in follow-up at pediatric cardiology. We observed that one in fifteen patients ceased follow-up completely; and one in four patients were untraceable. Regarding the setting and frequency of follow-up, the strictest interpretations of the recommendations (care within a formalized ACHD care program at particular timings) were implemented in one in four patients.

The study methodology was based on previous work of Goossens and coworkers allowing an indirect comparison of the present findings with those obtained in Belgium (14). Although our proportion of 'no cardiac follow-up' was comparable to that of the Belgian study (14), for about one quarter of patients no information on cardiac follow-up was, however, derived. This proportion of untraceability is substantially greater than found in the Goossens study (14), where only 1.1% was untraceable. It was even higher than the 12% untraceability in the study of Norris and colleagues, conducted in Cincinnati (OH) (20). These differences in proportions may be due to differences in migration rates. Indeed, emigration from the state of Massachusetts (2.4% in 2012) is substantially higher than that from Ohio (1.7% in 2012) or Belgium (0.8% in 2010) (21;22). This indicates that population and system variables should be taken into consideration when assessing continuity of care.

In the present study, we applied a four-step data collection strategy. Although the utilization of social media seems to be a trend-setting mean of communication that can be used in getting patients back on the radar for cardiac follow-up, IRB did not allow to contact patients through these websites for the purpose of this study (20;23). Although we cannot draw firm conclusions about the present cardiac care in untraceable patients, the prevalence of care gaps might be higher in these patients than in those who are contactable. Hence, patients with complex healthcare needs who disappear from the radar could experience discontinuous follow-up potentially leading to unrecognized complications, deterioration, or comorbidities and an impaired chance of receiving state-of-the-art care (24). Losing track of patients might be tackled by the development of nationwide databases, enabling professionals to keep track of patients within the overall healthcare system (25;26). Such systems might be of particular value during the transition phase and after

transferring patients to ACHD care, since this life event appeared to make patients vulnerable to care gaps (15). Our study is the first one identifying factors associated with untraceability. No or a lower number of surgeries in the past; health insurance issues; and a non-white ethnicity seemed to be risk factors for untraceability.

Although it is encouraging to observe that more than two-thirds of the patients received care in a specialized setting, analysis demonstrated that the majority of patients were not formally transferred to an adult-specific care setting, as three in four patients remained in pediatric cardiology at the age of 23y. This observation is fully in line with the Norris study (20), but is in large contrast with the Belgian study where only one in ten young adults was seen by pediatric cardiologists, either at the pediatric cardiology department or at a satellite center (14). This low proportion of adults seen by pediatric cardiologists in Belgium is due to a formal policy, agreed upon between pediatric and ACHD cardiologists, to transfer patients to adult care at the age of 16y. Indeed, institutional factors contributing to ongoing pediatric care were found to be an unlimited age access to pediatric care and a limited capacity of the adult clinic (20). Other barriers affecting transfer to ACHD care were the patients' unawareness of the need for follow-up, health insurance issues, not wanting further surgery, ambivalence towards the condition and not understanding the physician's explanation emotional or cognitive delay, non-compliance of patients with the transfer plan, unstable social situation or instability of the health condition (15;16;27). Oppositely, the most frequently indicated reasons for prompting transfer to ACHD care were life events such as pregnancy, marriage and school graduation, and not necessarily health problems such as adult comorbidities or health risk behaviors (28). In addition, in both the current study and that of Norris (25), the presence of an on-site ACHD care program within a pediatric medical facility had the potential ability to provide direct and indirect consultation to pediatric clinicians, influence pediatric-based care for adults with CHD, standardize care outcomes, and as such, may have lessened transfer of care to formalized ACHD care programs in young adults at an earlier age.

Overall, there was high implementation rate of the recommended frequency of follow up in both formalized ACHD care (94.2%) and pediatric cardiology (89%). However, only one quarter of patients were receiving care within a formalized ACHD care setting at the recommended frequency. Current recommendations advise a transfer to ACHD care when patients turn into adulthood (3;4;6;19;29;30). However, numerous influences to provision of ACHD care strictly within formalized care centers continue to exist in the U.S. as a model for national care delivery, including limited numbers of ACHD providers, emerging board certification for graduate medical education and development of competencies in ACHD care, patient and both medical and insurance provider knowledge of the need for (and allowance of the receipt of) specialty ACHD care, and both local and

regional established partnerships in effective care delivery. In addition, current ACHD guidelines are predominantly expert-based and lack vital empirical underpinning, which might explain the low implementation rates observed in our study. Future guidelines based on newly identified evidence that overall medical care provided in institutions that contain formalized specialty ACHD care does improve outcomes (11) and standardization of specialty ACHD care provision through board certification and program accreditation may provide the confidence that pediatric cardiologists require to routinely transfer adults to ACHD care within settings that contain a formalized ACHD care program.

S***tudy limitations***

This study was performed in one highly specialized pediatric hospital with both pediatric and ACHD services available for decades in a regional environment of more-widely accessible healthcare resources, limiting generalizability of results. Second, only patient-related characteristics were entered in the regression models predicting provision of care according to the recommendations and untraceability of patients.

Conclusion

We investigated the setting, type of clinician and frequency of outpatient visits received by young adults with CHD who were followed in pediatric cardiology during adolescence. Only one quarter of patients received care within formalized ACHD care at the frequency proposed by the guidelines. Retention to pediatric care was found to be high, indicating that transfer of care to an adult-oriented setting during young adulthood was not current standard practice. A considerable proportion of patients were untraceable in the healthcare system. Hence, this study demonstrated that additional efforts are needed to guarantee continuity of care across settings, time, and transitional life phases to patients with a chronic condition, such as CHD.

References

1. Marelli AJ, Ionescu-Ittu R, Mackie AS, Guo L, Dendukuri N, Kaouache M. Lifetime Prevalence of Congenital Heart Disease in the General Population from 2000 to 2010. Circulation 2014; 130(9):749-756.
2. van der Bom T, Zomer AC, Zwinderman AH, Meijboom FJ, Bouma BJ, Mulder BJ. The changing epidemiology of congenital heart disease. Nat Rev Cardiol 2011;8:50-60.
3. Sable C, Foster E, Uzark K, Bjornsen K, Canobbio MM, Connolly HM et al. on behalf of the American Heart Association Congenital Heart Defects Committee of the Council on Cardiovascular Disease in the Young, Council on Cardiovascular Nursing, Council on Clinical Cardiology, and Council on Peripheral Vascular DiseaseBest Practices in Managing Transition to Adulthood for Adolescents with Congenital Heart Disease: The Transition Process and Medical and Psychosocial Issues: A Scientific Statement From the American Heart Association. Circulation 2011;123:1454-1485.
4. American Academy of Pediatrics, American Academy of Family Physicians, American College of Physicians, Transitions Clinical Report Authoring Group. Supporting the health care transition from adolescence to adulthood in the medical home. Pediatrics 2011;128:182-200.
5. Mylotte D, Pilote L, Ionescu-Ittu R, Abrahamowicz M, Khairy P, Therrien J, et al. Specialized Adult Congenital Heart Disease Care: The Impact of Policy on Mortality. Circulation 2014;129:1804-1812.
6. Baumgartner H, Bonhoeffer P, De Groot NMS, de Haan F, Deanfield JE, Galie N, et al. Task Force on the Management of Grown-up Congenital Heart Disease of the European Society of Cardiology (ESC); Association for European Paediatric Cardiology (AEPC); ESC Committee for Practice Guidelines (CPG). ESC Guidelines for the management of grown-up congenital heart disease (new version 2010). Eur Heart J 2010;1-43.
7. Warnes CA, Liberthson R, Danielson GK, Dore A, Harris L, Hoffman JI et al. Task Force 1: The changing profile of congenital heart disease in adult life. J Am Coll Cardiol 2001;37:1170-1175.
8. Landzberg MJ, Murphy DJ, Davidson WR, Jarcho JA, Krumholz HM, Mayer JE Jr, et al. Task Force 4: Organization of Delivery Systems for Adults with Congenital Heart Disease. J Am Coll Cardiol 2001;37:1187-1193.
9. Yeung E, Kay J, Roosevelt GE, Brandon M, Yetman AT. Lapse of care as a predictor for morbidity in adults with congenital heart disease. Int J Cardiol 2008;125:62-65.
10. de Bono J, Freeman LJ. Aortic coarctation repair - lost and found: The role of local long term specialised care. Int J Cardiol 2005;104:176-183.
11. Wacker A, Kaemmerer H, Hollweck R, Hauser M, Deutsch MA, Brodherr-Heberlein S, et al. Outcome of operated and unoperated adults with congenital cardiac disease lost to follow-up for more than five years. Am J Cardiol 2005;95:776-779.
12. Reid GJ, Irvine MJ, McCrindle BW, Sananes R, Ritvo PG, Siu SC, et al. Prevalence and correlates of successful transfer from pediatric to adult health care among a cohort of young adults with complex congenital heart defects. Pediatrics 2004;113:e197-e205.
13. Mackie AS, Rempel GR, Rankin KN, Nicholas D, Magill-Evans J. Risk factors for loss to follow-up among children and young adults with congenital heart disease. Cardiol Young 2012;22:307-315.
14. Goossens E, Stephani I, Hilderson D, Gewillig M, Budts W, Van Deyk K, et al. SWITCH(2) Investigators. Transfer of Adolescents With Congenital Heart Disease From Pediatric Cardiology to Adult Health Care. An Analysis of Transfer Destinations. J Am Coll Cardiol 2011;57:2368-2374.
15. Gurvitz M, Valente AM, Borberg C, Cook S, Stout K, Kay J, et al. Prevalence and Predictors of Gaps in Care Among Adult Congenital Heart Disease Patients (The Health, Education and Access Research Trial: HEART-ACHD). J Am Coll Cardiol 2013;61:2180-2184.

16. Wray J, Frigiola A, Bull C, Adult Congenital Heart disease Research Network (ACoRN). Loss to specialist follow-up in congenital heart disease; out if sight, out of mind. Heart 2013;99:485-490.
17. Mitchell SC, Korones SB, Berendes HW. Congenital heart disease in 56,109 births. Incidence and natural history. Circulation 1971;43:323-332.
18. Vander Velde ET, Vriend JW, Mannens MM, Uiterwaal CS, Brand R, Mulder BJ. CONCOR, an initiative towards a nation registry and DNA-bank of patients with congenital heart disease in the Netherlands: rationale, design, and first results. Eur J Epidemiol 2005;20:549-557.
19. Warnes CA, Williams RG, Bashore TM, Child JS, Connolly HM, Dearani JA, del Nido P, Fasules JW, Graham TP Jr, Hijazi ZM, Hunt SA, King ME, Landzberg MJ, Miner PD, Radford MJ, Walsh EP, Webb GD, Smith SC Jr, et al.; American College of Cardiology; American Heart Association Task Force on Practice Guidelines (Writing Committee to Develop Guidelines on the Management of Adults With Congenital Heart Disease); American Society of Echocardiography; Heart Rhythm Society; International Society for Adult Congenital Heart Disease; Society for Cardiovascular Angiography and Interventions; Society of Thoracic Surgeons. ACC/AHA 2008 Guidelines for the Management of Adults With Congenital Heart Disease: A report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (Writing Committee to develop Guidelines on the Management of Adults with Congenital Heart Disease). Developed in Collaboration With the American Society of Echocardiography, Heart Rythm Society, International Society for Adult Congenital Heart Disease, Society for Cardiovascular Angiography and Interventions, and Society of thoracic Surgeons. Circulation 2008;118:714-833.
20. Norris MD, Webb GD, Drotar D, Lisec A, Pratt J, King E, et al. Prevalence and patterns of retention in cardiac care in young adults with congenital heart disease. J Pediatr 2013;163:902-904.
21. U.S. Census Bureau, 2012 American Community Survey. State-to-State Migration Flows: 2012 American Community Survey 1-Year Estimates. 2012. U.S.Census Bureau.
22. Algemene Directie Statistiek en Economische Informatie. Kerncijfers 2012. Statistisch overzicht van België. S000.01N/2012, 1-58. 2012. Brussels, Belgium, FOD Economie,KMO,Middenstand en Energie.
23. Valente AM, Lewis M, Vaziri SM, Bautista-Hernandez V, Harmon A, Kim Y, et al. Outcomes of Adolescents and Adults Undergoing Primary Fontan Procedure. Am J Cardiol 2013;112:1938-1942.
24. Iversen K, Vejlstrup NG, Sondergaard L, Nielsen OW. Screening of adults with congenital cardiac disease lost to follow-up. Cardiol Young 2007;17:601-608.
25. Oster ME, Riehle-Colarusso T, Simeone RM, Gurvitz M, Kaltman JR, McConnell M, et al. Public health science agenda for congenital heart defects: report from a Centers for Disease Control and Prevention Experts Meeting. J Am Heart Assoc. 2013; 2(5):e000256.
26. Wren C, O'Sullivan JJ. Loss to follow-up of adults with repaired congenital heart disease. Heart 2013;99:440-441.
27. Reiss JG, Gibson RW, Walker LR. Health care transition: youth, family, and provider perspectives. Pediatrics 2005;115:112-120.
28. Fernandes SM, Khairy P, Fishman L, Melvin P, O'Sullivan-Oliveira J, Sawicki GS, et al. Referral Patterns and Perceived Barriers to Adult Congenital Heart Disease Care. Results of a Survey of U.S. Pediatric Cardiologists. J Am Coll Cardiol 2012;60:2411-2418.
29. American Academy of Pediatrics, American Academy of Family Physicians and American College of Physicians-American Society of Internal Medicine. A consensus statement on health care transitions for young adults with special health care needs. Pediatrics 2002;110:1304-1306.
30. Foster E, Graham TP, Driscoll DJ, Reid GJ, Reiss JG, Russell IA, et al. Task Force 2: Special health care needs of adults with congenital heart disease. J Am Coll Cardiol 2001;37:1176-1183.

5 CURRENT EVIDENCE ON RISK AND PROTECTIVE FACTORS IMPACTING CARE GAPS IN YOUNG PEOPLE WITH COMPLEX CHRONIC CONDITIONS

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This chapter was submitted for review.

Abstract

Context: Care gaps are observed in an important proportion of young people transitioning to adulthood. Development of interventions preventing such drop-out of the medical system, as adolescents leave pediatric care, requires an understanding of the determinants of care gaps.

Objective: To systematically search the literature for determinants of care gaps in young people with chronic conditions transitioning to adulthood.

Data sources: Medline, Cinahl, and Embase were queried for peer-reviewed publications.

Study selection: Primary quantitative or mixed methods studies identifying determinants of care gaps in young people (10-25y) diagnosed with chronic conditions and written in English, French or Dutch were selected. Ten publications were included.

Data extraction: For each publication, the determinants of care gaps and quantitative results were extracted. Based on thematic analysis, determinants were categorized into four groups. Quantitative results were standardized, converting raw data into Odds Ratio's.

Results: Overall, 11 risk factors and nine protective factors for care gaps were identified in literature. All identified factors were related to the patient's characteristics. Demographics, disease-related characteristics, healthcare services use, and patient's health behaviors and beliefs were identified as significant determinants of care gaps in adolescents with chronic conditions.

Limitations: The large variability in study methods, statistical techniques and study populations resulted in inconsistent study findings.

Conclusions: This systematic review identified a set of patient-related determinants of care gaps. Unfortunately the internal and external validity of the study findings is limited, yielding the need for future prospective, multilevel studies addressing the remaining knowledge gaps

Introduction

As a result of improved medical therapies, diagnostic tools, and better survival rates for infants with life-threatening conditions, the prevalence of chronic health conditions in childhood increased substantially in Western countries [1;2]. The estimated prevalence was found to range from 3.5% to 35.3% [3]. An emerging population of special interest is the group of young people with complex chronic conditions (CCCs) defined as "*chronic physical, developmental, behavioral or emotional conditions lasting for at least 12 months, requiring specialized health services of a type or amount beyond that required by children in general*"[4-6]. CCCs represent a specific subgroup within the population of children diagnosed with a chronic condition, accounting for approximately 20% of this latter group [7-11]. The most frequent types of CCCs are of cardiovascular, congenital, neuromuscular, respiratory, or oncological nature[12]. In general, young people with CCCs account for a disproportionate use of healthcare resources due to their medical fragility. Indeed, these patients present with significantly higher hospitalization rates, readmissions, use of technology-dependent devices, prescribed medications, and inpatient mortality rates, yielding a financial and organizational burden for the current healthcare system [10-16].

Since most young people with CCCs are at risk for developing long-term complications, life-long specialized care is mandatory [13;17;18]. During childhood, care is generally provided at pediatric services, but as patients transition to adulthood, a transfer of care towards adult-oriented services is recommended [18]. This healthcare transition is an important but challenging life event [19] and a successful transfer to adult care is of paramount importance for young people with CCCs [20].

Studies reported that gaps in this transitional process are observed in 7-21% of young people with acromegaly [21-23], 9-17% of HIV-infected adolescents [24], 11-24% of patients with diabetes mellitus type I [25], and 7-76% of young people with congenital heart disease [26]. This drop-out of the medical system as adolescent patients leave pediatric care is associated with increased morbidity rates, long-term complications, increased number of hospitalizations, the need for urgent (re-)interventions, and higher rates of health-risk behaviors [22;25-35]. Therefore, measures to prevent such care gaps are of utmost importance.

In order to develop tailored preventive interventions or health care system reforms, a comprehensive understanding of the determinants of care gaps is required. Indeed, it can be assumed that not only patient-related factors predict care gaps, but also factors at the level of care organization within hospital, and factors related to the healthcare system at large. Since to date, no comprehensive overview of determinants of care gaps is available, a systematic review was

performed. The aim of this present review was therefore to systematically search the literature for determinants of care gaps in young people with CCCs transitioning to adulthood.

Methods

A systematic literature review was performed. The review and reporting are in line with the instructions of the 2009 PRISMA statement [36].

Search strategy

Three bibliographic databases, Medline, Cinahl, and Embase, were queried for peer-reviewed publications from inception to September 2014. A distinct search string was developed for each respective database (**see Table 5.1**) through subsequent brainstorm sessions, verified by a biomedical librarian, and supplemented with the snowball searching technique (i.e., screening of reference lists of relevant publications).

Eligibility criteria

Publications were selected if the following criteria were met: (i) primary studies including quantitative or mixed methods designs; (ii) studied sample comprised young people (aged 10-25y) diagnosed with (complex) chronic conditions[4-6] transferring to adult care; (iii) one of the aims of the study was to identify determinants of care gaps; (iv) written in English, French, or Dutch; and (v) published in peer-reviewed journals. Qualitative studies, editorials, comments, and letters to the editor were excluded. No restrictions with respect to the time of publication were, however, implemented.

A 'care gap' was defined as 'any type of discontinuation of the care process where the time lapse between mandatory follow-up visits exceeded the recommended period of time'. Care gaps comprised concepts such as lapse(s) of care [30], loss/lost to follow-up [37;38], unsuccessful transfer or transition [39-41], and cessation of follow-up [26].

Study selection

A total of 1,718 records were identified in the respective databases. After removal of 130 duplicates, the title and abstract of 1,588 records were screened using RefMan® software version 12.0 (Thomas Reuters). This screening was performed by two authors (EG, LB) independently. Finally, 8 full-text publications were selected for eligibility assessment by two authors (EG,LB) independently. At this stage, four publications were added using the snowball-technique. A total of 12 publications

were finally assessed for eligibility. Two publications were, however, excluded after a detailed review of the full-text; leaving ten publications to be included in this review (see **Figure 5.1**).

Data extraction

For each of the selected publications, a predetermined set of variables was extracted by one author (EG): first author; year of publication; setting; study design; method of data collection; sample characteristics; operational definition of the primary outcome; and statistical analyses. Furthermore, for each study, the determinants of care gaps and the quantitative results of the respective statistical tests were extracted. Results obtained through qualitative study designs or analysis techniques were, however, excluded. Based on a thematic analysis, we categorized determinants of care gaps into four groups: (i) demographic characteristics; (ii) disease-related characteristics; (iii) healthcare services use; and (iv) patient's behavior. Quantitative study results were standardized, converting reported p-values into Odds Ratio's (ORs) and 95% confidence intervals (CI), using a web-based calculator [42]. For some studies, however, insufficient data were reported enabling the calculation of these effect sizes.

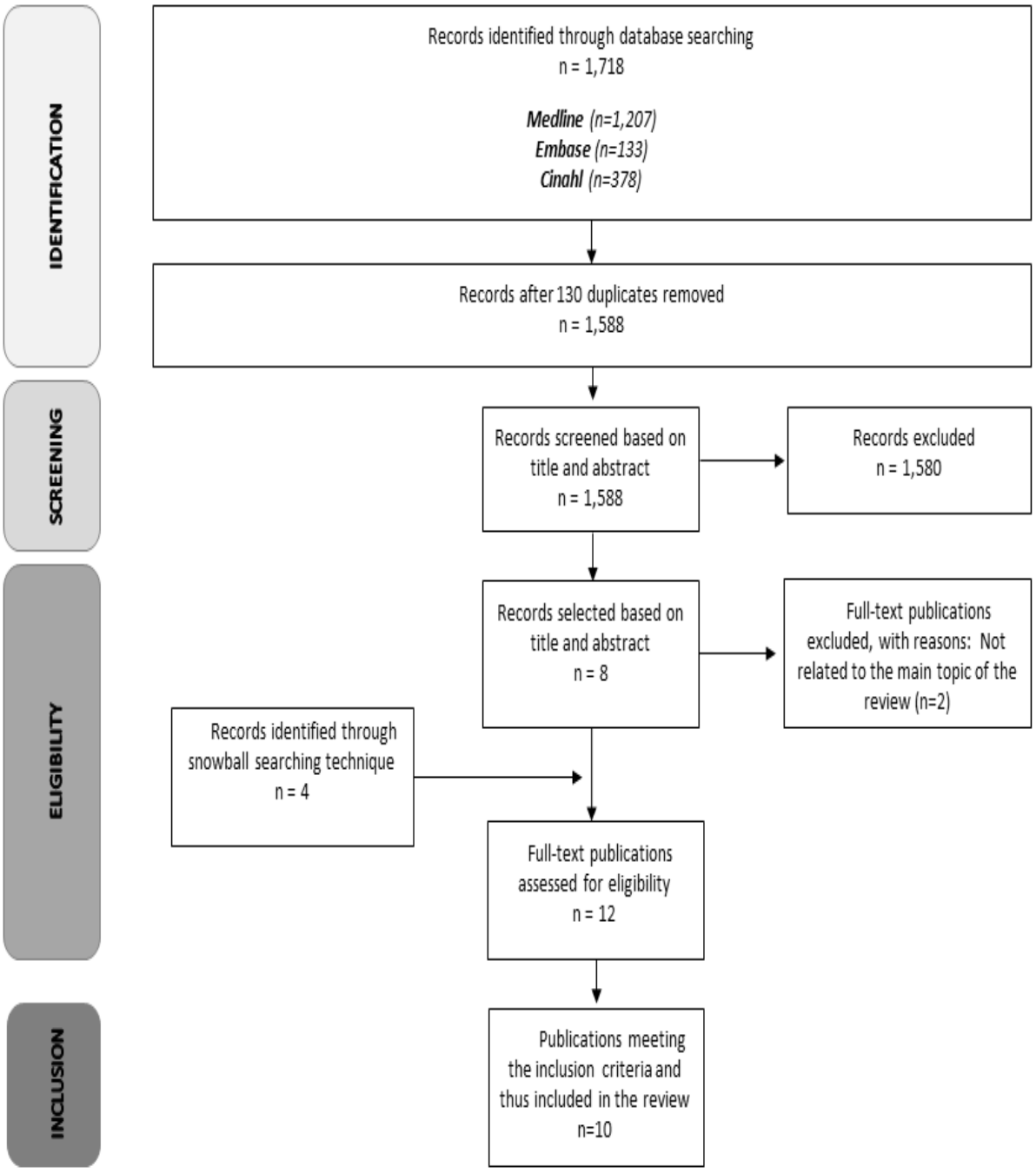
Assessment of the methodological study rigor

Since all selected studies had an observational non-randomized design, the methodological quality of every included article was critically appraised using an adapted version of the Methodological Index for Non-Randomized Studies (MINORS) instrument [43]. The original MINORS instrument comprises 12 items, of which the first eight items are applicable for non-comparative studies. The four remaining items are to be evaluated in case of a comparative study design. After critically revising all MINORS-items, we deleted two items since they were not applicable for our included studies. Items 5 ('unbiased (blind/double-blind) assessment of the study endpoint') and 7 ('attrition of study participants less than 5%') were found to be not applicable to the selected publications. Furthermore, although the original MINORS-instrument only assesses the appropriateness of applied statistical analysis techniques for comparative studies, we decided to evaluate this item for all studies. The respective MINORS-items were rated as 'not reported' (0), 'reported but inadequate' (1), or 'reported and adequate' (2)[43]. The use of this adapted MINORS-instrument, results in the calculation of a global score (maximum value of 14) for non-comparative studies and a global score for comparative studies (maximum value of 20) (**Table 5.2**).

Table 5.1: Overview of search strings

Medline	("Child"[Mesh:NoExp] OR child[tiab] OR child's[tiab] OR children[tiab] OR childhood[tiab] OR children's[tiab] OR kid OR kid's OR girl OR girls OR boy OR boys OR adolescents OR Adolescence OR teen OR teens OR teenager OR teenagers OR youth OR youths OR youngster*[tiab] OR adult child OR minors OR young adults OR young adul* OR emerging adul* OR junior high OR middle-school OR high-school OR juveniles OR "Pediatrics"[Mesh:NoExp] OR Pediatrics[tiab] OR Pediatric[tiab] AND (chronic diseases OR Chronic Illness* OR Chronically Ill OR persistent illness* OR persistent disease[tiab] OR long term illness* OR long term disease* OR special health need*[tiab] OR special healthcare need*[tiab] OR special health care need*[tiab] OR "multiple disabilities" OR systemic disorder[tiab] OR systemic disease [tiab] OR heritable disease[tiab] OR heritable disorder [tiab] OR heritable condition [tiab] OR chronic health condition [tiab] OR lifelong disability[tiab] OR life long disability [tiab] OR "genetic diseases, inborn"[MeSH Terms] OR Inborn Genetic Diseas* OR Hereditary Diseas* OR "congenital, hereditary, and neonatal diseases and abnormalities"[MeSH Terms] OR Congenital Disorder* OR neonatal diseas* OR Developmental disability) AND (continuity of patient care[tiab] OR "transition to adult care" OR "Care Continuum" OR "Care Continuity" OR "loss to follow up" OR "lost to follow-up" OR "lost to follow up" OR "care gap" OR "transfer of care" OR "transition to adult care" OR healthcare transition*[tiab] OR health care transition*[tiab] OR "lifelong care" OR "life long follow-up" OR "lifelong follow-up" OR "continuity of care" OR "transitional care" OR "successful transfer") AND ((english[Language]) OR french[Language]) OR dutch[Language]) NOT (((editorial [Publication Type]) OR comment[Publication Type]) OR letter[Publication Type]))
Embase	('child' OR 'child':ti OR 'child':ab OR 'children':ti OR 'children':ab OR 'childhood':ti OR 'childhood':ab OR kid OR girl OR boy OR girls OR boys OR adolescents OR adolescence OR teen OR teens OR teenager OR teenagers OR youth OR youths OR minors OR young NEXT/1 adult OR young adults OR young adulthood OR youngster*:ti OR youngster*:ab OR adult child OR junior high OR 'middle school' OR 'middle-school' OR 'high-school' OR juvenile OR juveniles OR pediatrics OR 'pediatrics'/de OR 'pediatric':ti OR 'pediatric':ab OR 'pediatrics':ti OR 'pediatrics':ab) AND ('chronic disease'/exp OR chronic illness* OR chronically ill OR persistent illness* OR 'persistent disease':ti OR 'persistent disease':ab OR (long AND term illness*) OR (long AND term AND diseas*) OR (special AND health AND care AND need*:ti) OR (special AND health AND need*:ab) OR (special AND health AND need*:ti) OR (special AND healthcare AND need*:ti) OR (special AND healthcare AND need*:ab) OR multiple disabilities OR 'systemic disorder':ti OR 'systemic disorder':ab OR 'systemic disease':ti OR 'systemic disease':ab OR 'heritable disease':ti OR 'heritable disease':ab OR 'heritable disorder':ti OR 'heritable disorder':ab OR 'heritable condition':ti OR 'heritable condition':ab OR 'chronic disease'/exp OR 'chronic health condition':ti OR 'chronic health condition':ab OR 'lifelong disability':ti OR 'lifelong disability':ab OR 'life long disability':ti OR 'life long disability':ab OR 'genetic disorder'/de OR (inborn AND genetic AND diseas*) OR (hereditary AND diseas*) OR 'congenital disorder'/exp OR congenital disorder* OR congenital diseas* OR neonatal diseas* OR developmental disability) AND ((continuity AND of AND patient AND care) OR (continuity care) OR 'transition to adult care'/exp OR 'patient care'/de OR care continuum OR care continuity OR (loss AND to AND 'follow up':ti) OR (loss AND to AND 'follow up':ab) OR (lost AND to AND 'follow up':ti) OR (lost AND to AND 'follow up':ab) OR (loss AND to AND 'follow-up':ab) OR (lost AND to AND 'follow-up':ti) OR care gap OR transfer care OR 'transfer of care' OR 'healthcare transition' OR healthcare transition OR lifelong care OR (life AND long care) OR (life AND long 'follow up') OR lifelong 'follow up' OR continuity care OR continuity care OR transitional care OR successful transfer) AND ([dutch]/lim OR [english]/lim OR [french]/lim) AND ([article]/lim OR [article in press]/lim OR [conference abstract]/lim OR [conference paper]/lim OR [conference review]/lim OR [review]/lim)
Cinahl	((((((((english[Language]) OR french[Language]) OR dutch[Language])))) AND ((((continuity of patient care[tiab] OR "transition to adult care" OR "Care Continuum" OR "Care Continuity" OR "loss to follow up" OR "loss to follow-up" OR "lost to follow up" OR "lost to follow-up" OR "care gap" OR "transfer of care" OR "transition to adult care" OR healthcare transition*[tiab] OR health care transition*[tiab] OR "lifelong care" OR "life long follow-up" OR "lifelong follow-up" OR "continuity of care" OR "transitional care" OR "successful transfer")) AND ((("Child"[Mesh:NoExp] OR child[tiab] OR child's[tiab] OR children[tiab] OR childhood[tiab] OR children's[tiab] OR kid OR kid's OR girl OR girls OR boy OR boys OR adolescents OR Adolescence OR teen OR teens OR teenager OR teenagers OR youth OR youths OR youngster*[tiab] OR adult child OR minor OR young adults OR young adul* OR emerging adul* OR junior high OR middle-school OR high-school OR juveniles OR "Pediatrics"[Mesh:NoExp] OR Pediatrics[tiab])) AND ((chronic diseases OR Chronic Illness* OR Chronically Ill OR persistent illness* OR special health need*[tiab] OR special healthcare need*[tiab] OR special health care need*[tiab] OR "multiple disabilities" OR systemic disorder[tiab] OR heritable condition [tiab] OR heritable disorder [tiab] OR heritable disease[tiab] OR systemic disease [tiab] OR lifelong disability[tiab] OR life long disability [tiab] OR "genetic diseases, inborn"[MeSH Terms] OR Inborn Genetic Diseas* OR Hereditary Diseas* OR "congenital, hereditary, and neonatal diseases and abnormalities"[MeSH Terms] OR Congenital Disorder* OR neonatal diseas* OR Developmental disabilities))))) NOT ((((editorial[Publication Type]) OR comment[Publication Type]) OR letter[Publication Type]))))

Figure 5.1: Flow diagram of the study selection process



Results

Study characteristics

Within the ten selected publications, four type of CCCs were studied: sickle cell disease (SCD) [39;44], congenital heart disease (CHD) [26;30;38;40;45;46], congenital adrenal hyperplasia (CAH) [37], and juvenile idiopathic arthritis (JIA) [41]. Two of these studies explicitly stated that patients diagnosed with a neurodevelopmental condition were excluded [40;46]. The majority of studies (80%) were performed in the USA [30;39;40;44;46] or Canada [38;41;45], while only two studies were conducted in Europe[26;37]. Sample sizes ranged from 18 to 922 young people (median 227). Although most studies did not report on the ethnic composition of their sample [30;37;38;40;41;45], the majority of studied samples were either exclusively African-American [39;44] or White, Caucasian [26;41].

Seven studies used a retrospective descriptive design [26;30;37;39;41;44;45], two studies used a cross-sectional descriptive design [40;46], and one study used a limited mixed methods approach combining a matched case-control quantitative study with a qualitative interview study[38]. Most studies (70%) collected both data regarding care gaps as well as data on potential determinants based on a retrospective review of medical records or clinical databases [26;37-41;44]. Some other studies used self-reported methods such as surveys, telephone interviews or structured face-to-face interviews to collect these data [26;30;38;40;44;46]. One study analyzed a province-wide administrative database [45]. Single center studies were most represented [26;30;37-39;41], although four studies used a multicentric study design [40;44-46] (**Table 5.3**).

Quality assessment of research methodology

The MINORS global quality score ranged from 15 to 16 out of 20 for comparative studies [38;39], and from 7 to 12 out of 14 for non-comparative studies [26;30;37;40;41;44-46]. With the exception of one study [44], all eligible patients were included, follow-up periods were adequate, and a clear explanation was provided of the criteria used to evaluate the outcome of care gaps. Eight studies (80%) collected data on determinants retrospectively [26;37-39;41;44;45;47], and none of the studies performed a power analysis with a prospective calculation of the required sample size. The statistical analyses identifying determinants of care gaps were adequate in six (60%) studies [26;30;38;40;45;46] using multivariate logistic regression analyses accounting for potential confounders. Four studies, however, only performed two-group comparisons[37;39]; did not correct for multiple testing[37;39;41;44]; calculated correlation coefficients[44], or performed only univariate analysis[41] (**Table 5.2**).

Table 5.2: Critical appraisal of methodological rigor using adapted MINORS-tool

	Wojciechowski et al. (2002)[44]	Reid et al. (2004)[40]	Yeung et al. (2008)[30]	Mackie et al. (2009)[45]	Hazel et al. (2010)[41]	Goossens et al. (2011)[36]	Mackie et al. (2012)[38]	Gleeson et al. (2013)[37]	Gurvitz et al. (2013)[46]	Andemariam et al. (2014)[39]
(Non-)comparative studies										
1. Clearly stated aim	2	2	2	2	2	2	2	2	2	2
2. Inclusion of consecutive patients: all eligible patients were included during the study period	1	2	2	2	2	2	2	2	2	2
3. Data collection: data collection according to a protocol established before the beginning of the study	1	2	2	1	1	1	1	1	2	1
4. Endpoints appropriate to the aim of the study: unambiguous explanation of criteria used to evaluate the main outcome	1	2	2	2	2	2	2	2	2	2
5. Follow-up period appropriate to the aim of the study: follow-up period should be sufficiently long allowing assessment of main outcome	1	2	2	2	2	2	2	2	2	2
6. Prospective calculation of the study size: power calculation and provision of 95% confidence interval, according to the expected incidence of the outcome event	0	0	0	0	0	0	0	0	0	0
7. Adequate statistical analysis: statistics in accordance with the study design and calculation of confidence intervals or relative risk	1	2	2	2	1	2	2	1	2	1
Comparative studies										
8. Adequate control group	N/A	N/A	N/A	N/A	N/A	N/A	1	N/A	N/A	2
9. Contemporary groups: control and studied group managed during the same time period (no historical comparison)	N/A	N/A	N/A	N/A	N/A	N/A	2	N/A	N/A	1
10. Baseline equivalence of groups: groups are similar regarding criteria other than studied endpoints	N/A	N/A	N/A	N/A	N/A	N/A	2	N/A	N/A	2
MINORS global score	7/14	12/14	12/14	11/14	10/14	11/14	16/20	10/14	12/14	15/20

The items are scored 0 (not reported); 1 (reported but inadequate); or 2 (reported and adequate). The global score being 14 for non-comparative and 20 for comparative studies, respectively.

Table 5.3: Overview of study characteristics

Reference	Wojcie-chowski et al. (2002)[44]	Reid et al. (2004)[40]	Yeung et al. (2008)[30]	Mackie et al. (2009)[45]	Hazel et al. (2010)[41]	Goossens et al. (2011)[26]	Mackie et al. (2012)[38]	Gleeson et al. (2013)[37]	Gurvitz M. et al. (2013)[46]	Andemariam et al. (2014)[39]
Study design	Descriptive study	Cross-sectional cohort study	Descriptive study	Descriptive study	Descriptive study	Descriptive study	Mixed methods study including matched case-control study	Descriptive study	Descriptive study	Descriptive study
Country	USA	Canada	USA	Canada	Canada	UK	Canada	UK	USA	USA
Setting	Multicentric; 3 centres	Multicentric; 15 centres	Singlecentre	Province-wide (Québec)	Singlecentre	Singlecentre	Singlecentre	Singlecentre	Multicentric; 12 centres	Singlecentre
Method of data collection	Retrospective chart review & structured interviews	Retrospective review of clinical database & self-administered questionnaire & structured interviews	Interviews	Retrospective review of province-wide database	Retrospective chart review	Retrospective review of clinical data, chart review & electronic appointment system	Retrospective chart review & interviews	Retrospective review of clinical data, chart review & electronic appointment system	Survey	Retrospective chart review
Type of CCC	SCD	CHD	CHD	CHD	JIA	CAH	CHD	CAH	CHD	SCD
Sample size	n=18	n= 360	n=158	n=643	n= 100	n=61	n=296	n=61	n=922	n=47
Proportion women	50%	45%	66%	48%	68%	44%	37,5%	44%	54%	57%
Ethnicity	100% African-American	Not reported	Not reported	Not reported	Not reported	Not reported	Not reported	Not reported	83% Caucasian	87% African – American

CAH = Congenital Adrenal Hyperplasia; CCC = Complex Chronic Condition; CHD = Congenital Heart Disease; JIA = Juvenile Idiopathic Arthritis; SCD = Sickle Cell Disease; UK = United Kingdom; USA = United States of America

Table 5.4: Operational definitions of care gaps

First author (year of publication)	Concept	Definitions	Setting of care	Time interval	Data source
Wojciechowski (2002)[44]	Uninterrupted care	Patients follow through with the adult-centered care post-transfer; measured by whether or not patients kept their initial adult appointment and the time between last pediatric and first subsequent adult visit.	adult-centered SCD care ‖	not specified †	retrospective chart review
Reid(2004) [40]	Successful transfer	Attendance at an ACHD center, ≥1 appointment of any type (ECG, outpatient visit, echocardiography, …)	ACHD center *	1 year ‖	ACHD centers database and self-report survey
Yeung(2008)[30]	Lapse in care	≥2 years interval from leaving pediatric cardiac care facility and attending ACHD care.	ACHD care *	≥2 years *	clinic database
Mackie(2009)[45]	Lack of cardiology follow-up	Lack of an outpatient assessment by a cardiologist or attrition at a specific age group.	any type of cardiologist, both specialized as non-specialized in CHD *	attrition per age group, years in between age groups ‖	province-wide administrative database
Hazel(2010)[41]	Unsuccessful transfer	Failure to continue follow-up with an adult rheumatologist 2 years after transfer	specialized adult care *	2 years post-transfer *	chart review
Goossens(2011) [26]	No follow-up	Patient indicated he/she was currently not in cardiac follow-up or patient was untraceable.	in cardiac follow-up, settings could vary from specialist cardiology to shared care to non-specialist cardiac care (GP excluded) ‖	3 year post-transfer ‖	clinical database or patient self-report
Mackie(2012)[33]	Lost to follow-up	No return visit to cardiac clinic for period of ≥3 years.	cardiac clinic (any type of cardiologist) *	≥3 year post-transfer *	retrospective chart review
Gleeson (2013)[37]	Lost to follow-up	Patient stopped attending pediatric or adult specialized clinic or was discharged from pediatric care because of non-attendance to care before transfer to adult care could occur.	specialized CAH clinic *	3 year post-transfer ‖	clinical data and electronic appointment system
Gurvitz(2013)[46]	Care gap	>3 years gap in cardiology care	any type of cardiac care *	>3 years *	patient self-report
Andemariam (2014)[39]	Unsuccessful transition	No attendance of at least 1 outpatient visit at adult SCD clinic 1 year after transfer from pediatric clinic.	adult SCD clinic *	1 year post-transfer *	retrospective chart review

** Specified = clearly stated in the operational definition; ‖ Indirectly specified = not clearly stated in operational definition but deductible from the study methodology; † Not specified = not stated, nor described or deductible from study methodology; SCD = Sickle Cell Disease; ACHD = Adult Congenital Heart Disease; GP = General Practitioner*

Definitions for care gaps

A broad range of definitions of care concepts has been provided in the selected publications (**Table 5.4**). All studies provided an operational definition in their manuscript. However, the level of specification of the setting, time interval, and data source varied largely across studies. Some studies provided highly detailed information on these components, while for others; specifications could indirectly be derived from the study methodology or result section. Only one study did not provide any kind of information on the time interval considered for the determination of a care gap [44]. Detailed information on the concepts, operational definitions, and specifications is provided as **Table 5.4**.

Determinants of care gaps

A total of 45 potential determinants of care gaps have been investigated. Either factors that could increase or decrease the likelihood of experiencing care gaps were identified. These factors can be considered as 'risk factors' or 'protective factors', respectively. Overall, 11 risk factors and nine protective factors were identified. An overview of determinants of care gaps identified is provided four tables (**Tables 5.1-5.4**).

Demographics

Living independently from parents [30], male sex [26;45], lower family income [38], and longer travel distance to closest adult specialized clinic [37;39] were identified as significant risk factors, although for the latter three factors non-significant results were reported as well. Older age at last pediatric visit was inconsistently found to be a risk factor [39] and a protective factor[40]. Ethnicity [39;44;46]; type of healthcare insurance[39;44]; type of residence (urban/rural)[45]; age at diagnosis[30]; and educational level[41;46] were not significantly related to care gaps (**Table 5.5**).

Table 5.5: Demographical determinants of care gaps

DEMOGRAPHICS			
	Significant risk factors	Significant protective factors	Non-significant results reported
Living independently from parents	OR=4.1; 95%CI 1.7-10.1[30]		
Male sex	OR=1.80; 95%CI 1.02-3.17[26]		OR=0.57; 95%CI 0.57-1.23[39]
	OR=1.52; 95%CI 1.05-2.20[45]		OR=1.23; 95%CI 0.61-1.98[37]
			OR=2.15; 95%CI 0.91-2.61[41]
			[44] ‡[46]‡
Higher family income		OR=.87 per increase in annual salary of 10,000 $; 95%CI .77-.98[38]	[40]‡
Older age at last pediatric visit	OR=10; 95%CI 1.71-58.43[39]	OR=1.29; 95%CI 1.10-1.51[40]	
Travel distance to closest adult specialized centre	OR= 7.71; 95%CI 1.53-38.83)[39]		OR=0.56; 95%CI 0.27-1.17[40]
	p=.03 ($\bar{x}$ 14 versus 24 miles)[37]‡		p=0.34 [38]‡; [26]‡
Ethnicity			p=.67 [39]‡; [44]‡
Type of healthcare insurance			OR=0.51; 95%CI 0.56-1.21[39]; [44]‡
Type of residence (urban, rural)			[45]‡
Age at diagnosis			OR=1; 95%CI 1.0-1.1 [30]
Educational level			OR=0.49; 95%CI0.46-1.08 [41]; [46]‡

‡ *Insufficient data were reported to convert raw data into Odds Ratio's (Ors) and 95% Confidence Intervals (CIs).*

Disease-related characteristics

Milder disease activity, severity or complexity was a significant risk factor for care gaps [30;37;39;41;45;46]. The study of Goossens and colleagues, however, did not find a significant impact of disease complexity on care gaps [26]. For patients with CHD, not having undergone heart surgery in the past also increased the risk of having care gaps [26]. Conversely, having at least one comorbid condition was found to be protective for the occurrence of care gaps [40]. Determinants which were investigated but found to be non-significant were: residual hemodynamic problems at last echocardiography, medication use, and an implanted pacemaker or cardio defibrillator [38] (**Table 5.6**).

Healthcare services use

Regarding the use of healthcare services, a lower number of outpatient visits in pediatric care during the last three years pre-transfer [37;45]; last visit organized outside a university hospital [45]; hospitalizations in childhood [45]; and a history of at least one missed appointment [38] were significant risk factors for care gaps. Furthermore, the multicentric study of Gurvitz et al. (2013) found a significant difference between locations of hospitals (i.e., Colorado, Oregon, Washington State) in the occurrence of care gaps. In contrast, having a written recommendation on the type of professional that should provide adult follow-up care[38]; and adherence to the first or second outpatient visit in adult clinic (i.e. good early attenders)[37;44] were identified as protective factors (**Table 5.7**).

Table 5.6: Disease-related determinants of care gaps

DISEASE-RELATED CHARACTERISTICS			
	Significant risk factors	Significant protective factors	Non-significant results reported
Milder disease severity, complexity or activity	OR= 0.26; 95%CI 0.07-0.95[39]		[26]‡
	OR= 4.31 ; 95%CI 0.77-24,14[37]		
	OR= 4.14; 95%CI 2.17-7.87[45]		
	OR= 2.3; 95%CI 1.2-4.5[30]		
	OR=2.67; 95%CI 1.16-6.16[41]		
	OR=2.2 to 4.1, p<.0001[46]		
≥1 co-morbid condition		OR=3.13; 95%CI 1.13-8.67[40]	OR=1.12; 95%CI 0.62-2.04[38]
No heart surgery performed in the past (for CHD)	OR=5.97; 95%CI 3.04-11.72[26]		
Residual hemodynamic problems at last echo-cardiography			OR=1.69; 95%CI 0.93-3.07[38]
Medication use			OR=1.48; 95%CI 0.84-2.59[38]
Implanted pacemaker or automatic implantable cardiac defibrillator			OR= 8.00; 95%CI 0.84-75.81[38]

‡ *Insufficient data were reported to convert raw data into Odds Ratio's (Ors) and 95% Confidence Intervals (CIs).*

Table 5.7: Determinants of care gaps related to healthcare services use

	HEALTHCARE SERVICES USE			
	Significant risk factors	Significant protective factors	Non-significant results reported	
Lower number of pediatric out-patient visits during pre-transfer period	p=.01‡ [37]; OR=1.15 per 1 visit decrease; 95%CI 1.09-1.21[45]			
Last follow-up visit outside of university hospital	OR=1.63; 95%CI 1.02-2.61[45]		OR=2.09; 95%CI 0.71-6.13 [38]	
Non-cardiac related hospitalizations	OR=1.93; 95%CI 1.18-3.17[45]			
Cardiac related hospitalizations without invasive procedures	OR=2.22; 95%CI 1.36-3.62[45]			
History of ≥ 1 missed appointment	OR=13.0; 95%CI 3.3-51.7[38]			
Geographical location of clinic	p<.001 (Colorado); p<.002 (Oregon); p<.027 (Washington State) [46‡]			
Written recommendation on type of professional performing adult care		OR= 0.4; 95%CI 0.2-0.8[38]		
Keeping 1st & 2nd appointment in adult care (i.e., good early attenders)		OR= 5.2; 95%CI 1.25-21.57 [37];p<.05‡ [44]		

‡ Insufficient data were reported to convert raw data into Odds Ratio's (Ors) and 95% Confidence Intervals (CIs).

Patient's behavior

Greater independence in attending appointments [40]; patient's belief that follow-up should be continued in specialized adult care[40]; higher levels of self-efficacy [44]; abstaining from substance use and full compliance with antibiotic prophylaxis regimens[40] were identified as significant protectors for care gaps. Non-significant determinants were: self-reported family functioning[40], health beliefs [40], SF-36 scores[40], self-rated activity restictions[40], general preferences of self-care[40], patient-reported expected frequency of visits in adult clinic[40], treatment adherence[44], and patients' knowledge of disease name[46] (**Table 5.8**).

Table 5.8: Determinants of care gaps related to patient's behavior

PATIENT'S BEHAVIOR			
	Significant risk factors	Significant protective factors	Non-significant results reported
Patients' belief that follow-up should be performed at specialized adult clinic		OR=3.64; 95%CI 1.34-9.9[40]	
No substance use		OR=.18; 95%CI .07-.50[40]	
Compliance to antibiotic prophylaxis		OR=4.23; 95%CI 1.48-12.06[40]	
Greater independence at attending appointments		OR=6.59; 95%CI 1.61-27 [40]	
Higher levels of self-efficacy		p<.05‡ [44]	
Self-reported family functioning			[40]‡
Health beliefs			[40]‡
SF-36 scores			[40]‡
Self-rated activity restrictions			[40]‡
General preferences for self-care			[40]‡
Patient-reported expected frequency of visits to adult clinic			[40]‡
Treatment adherence			[44]‡
Patient's knowledge of defect's name			[46]‡

‡ Insufficient data were reported to convert raw data into Odds Ratio's (Ors) and 95% Confidence Intervals (CIs).

Discussion

Over the past decades, survival rates improved substantially for children diagnosed with a complex chronic condition (CCC) resulting in better prospects towards achieving adult life[1;2]. However, in order to guarantee that improved survival rates result in improved quality of life, enhanced health status, and prevention of complications, the provision of continuous care is mandatory. International guidelines stress the importance of providing uninterrupted, age- and developmentally appropriate health care to patients with CCCs throughout their life span [13;17;18]. Safe-guarding continuity of care appears, however, challenging in adolescents and young adults diagnosed with CCCs. Adolescence is the typical life phase in which authority is challenged and risk-taking behaviors, possessing additional risks for patients with CCCs, are more prominent [48;49]. These developmental changes, in combination with the required transfer of healthcare setting, embody an important challenge for adolescents with CCCs. Previous studies demonstrated that an important proportion of young people with CCCs experience care gaps, or are even completely lost to follow-up [21-26].

Improving continuity of care for young people with CCCs, requires tailored interventions or strategies facilitating a smooth and continuous care process. An in-depth understanding of determinants of care gaps is, nevertheless, indispensable for the development of such interventions. Since a comprehensive overview of determinants of care gaps is currently missing, a systematic review of the existing literature was performed. This review identified ten relevant publications that were performed in young people diagnosed with a complex chronic condition. Four types of CCCs were represented: sickle cell disease[39;44], congenital adrenal hyperplasia [37], congenital heart disease [26;30;38;40;45;46], and juvenile idiopathic arthritis[41]. No other types of CCCs have been investigated. The issue of care gaps appeared to be most often studied in a population of young people with congenital heart disease[26;30;38;40;45;46].

A total of 11 risk factors and nine protective factors were identified. Based on a thematic analysis, these determinants could be categorized in four different group: (i) demographics, (ii) disease-related characteristics, (iii) healthcare services use, and (iv) patient's behavior.

Milder disease activity, severity, or complexity was the only risk factor for which all studies analyzing this disease-related characteristic, unanimously reported significant results [30;37;39;41;45;46]. Furthermore, having no or less comorbid conditions [40] and not having undergone heart surgery in the past [26], both indirect indicators of milder disease, were significant determinants of care gaps. Hence, special attention should be paid to patients diagnosed with a milder type of CCC since these patients

seem to have an increased likelihood of experiencing care gaps. These patients might perceive their long-term risk for mortality and morbidity to be low as compared to patients with moderate-to-complex conditions. Tailored interventions aiming to convince patients of the benefits of continued follow-up care could be implemented in practice as a preventive measure of care gaps.

Demographical characteristics such as male sex[26;45], family income[38], age at last pediatric visit[39], travel distance[37;39], and living independently from parents[30] were identified as significant determinants of care gaps. Unfortunately, non-significant results were also reported for some of these demographical risk factors leading to inconsistent research findings. These patient-related characteristics are less modifiable but could be used in practice when screening for patients at risk for care gaps. Finally, more hospitalizations[45], less outpatient visits during childhood[45], and a history of missed appointments[38] increased the risk for care gaps while patients who demonstrated better self-management skills were less likely to experience care gaps [40;44] .

Overall, this review revealed that a rather limited set of patient-related characteristics is known to alter the risk for care gaps in young people with CCCs transitioning to adult care. Most factors were related to demographics, characteristics of the disease, or the healthcare use of patients. Although most identified risk factors are modifiable to a limited extent, some protective factors such as guaranteeing that patients leave pediatric care with a written recommendation on the type of professional providing follow-up adult care, or checking if patients attend the first outpatient visits in adult care, might be components of interventions preventing care gaps.

Although this review aimed to identify determinants of care gaps, no uniform definition for the concept of care gaps currently exists. This review revealed a large heterogeneity of operational definitions provided for the concept of discontinuation of care. Synonyms used for care gaps[46] were: uninterrupted care[44], lapse in care [30], lack of follow-up [45], no follow-up [26], lost to follow-up [37;38], and unsuccessful transition [39]. All studies provided a relatively detailed operational definition, specifying which setting of care and respective time interval in-between visits were evaluated when determining the occurrence of care gaps. In order to enhance comparability of study results in the future, consensus should be reached on the operational definition of care gaps. Additional efforts should be made to provide such a uniform definition for this concept.

Although all studies aimed to identify determinants of care gaps, a wide variety of statistical techniques, research designs, and data collection methods were used to address this research aim. Determinants were identified through a variety of techniques such as: two-group comparisons, calculation of correlation coefficients, or uni- and multivariable regression analysis. From a

methodological point of view, there is an important difference in the robustness of results obtained through the performance of repeated group comparisons without correction for multiple testing versus multivariate regression analyses. Furthermore, critical appraisal of the methodological rigor of the included publications revealed that most studies analyzed determinants in a retrospective manner. Retrospective analyses, however, are characterized by the risk of missing information on specific factors not included in the previously established dataset.

Despite the transparent, objective, and rigorous nature of this review, some methodological limitations should be kept in mind. First, although relevant literature was searched by using a rigorously developed search string, only 10 out of 1,718 (<0.5%) retrieved records appeared to be relevant. The inclusion of the search term 'loss/lost to follow-up' most likely resulted in a large number of unrelated publications. This search term is very often used in the abstract of papers, within the context of attrition of participants from a longitudinal study. This search term was, however, indispensable for our search string since it is often used as a synonym for a care gap. Furthermore, additional relevant references might have been overlooked when indexed in other databases or only retrievable in grey literature. Secondly, the development of forest plots or the performance of a meta-analysis would have been of high value when investigating determinants of care gaps. Unfortunately, these analysis techniques could not be applied due to a large variability in study designs, data collection methods, sample sizes, low number of studies investigating a specific factor and the lack of sufficient raw data. Thirdly, generalizability of study results is fairly low since only four types of CCCs were investigated; patients with neurodevelopmental impairments were explicitly excluded in most studies; and sample sizes were small to moderate.

Although this systematic review identified factors associated with care gaps, additional research projects are needed to address the remaining knowledge gaps. Prospective, multicentric, international study designs analyzing both clinical and administrative datasets are highly needed to explore the relationship between care gaps and patient-, hospital-, and healthcare system-related factors. Furthermore, in order to enlarge the internal and external validity of research findings summarized in this review, additional studies should be performed in larger patient populations diagnosed with other types of CCCs.

Conclusion

This systematic literature review identified a set of determinants related to the characteristics of young people with CCCs found to increase or decrease the risk of experiencing a care gap. All identified determinants were related to the individual patient reflecting demographics, disease-

related characteristics, the use of healthcare services, and patient's behavior. Previous studies demonstrated that the transition to adulthood is a vulnerable period for young people with CCCs characterized by care gaps. Strategies to prevent such gaps in the care process are highly needed but additional prospective, multicentric, international research projects seem to be mandatory to enlarge our body of evidence. Furthermore, a uniform operational definition of a care gap is needed in order to increase the comparability of study findings.

References

(1) World Health Organization. Global status report on noncommunicable diseases 2010. Alwan A, editor. 1-176. 2010.

(2) Mladovsky P, Allin S, Masseria C, Hernàndez-Quevedo C, McDaid D, Mossiabos E. Health in the European Union. Trends and analysis. Observatory Studies Series N°19, 1-200. 2009. World Health Organization, on behalf of the European Observatory on Health Systems and Policies.

(3) van der Lee JH, Mokkink LB, Grootenhuis MA, Heymans HS, Offringa M. Definitions and Measurement of Chronic Health Conditions in Childhood. JAMA 2007;297:2741-2751.

(4) McPherson M, Arango P, Fox H et al. A new definition of children with special health care needs. Pediatrics 1998;102:137-140.

(5) Feudtner C, Christakis DA, Connell FA. Pediatric deaths attributable to complex chronic conditions: a population-based study of Washington State, 1980-1997. Pediatrics 2000;206:205-209.

(6) Pewacheck PW, Strickland B, Shonkoff JP et al. An epidemiologic profile of children with special health care needs. Pediatrics 1998;102:117-123.

(7) Perrin JM, Bloom SR, Gortmaker SL. The increase of childhood chronic conditions in the United States. JAMA 2007;297:2755-2759.

(8) Kenney GM, Ruhter J, Selden TM. Containing costs and improving care for children in Medicaid and CHIP. Health Aff (Millwood) 2009;28:1025-1036.

(9) Liptak GS, Shone LP, Auinger P, Dick AW, Ryan SA, Szilagyi PG. Short-term persistence of high health care costs in a nationally representative sample of children. Pediatrics 2006;118:e1001-e1009.

(10) van Dyck PC, Kogan MD, McPherson M, Weissman GR, Newacheck PW. Prevalence and characteristics of children with special health care needs. Arch Pediatr Adolesc Med 2004;158:884-890.

(11) Child and Adolescent Health Measurement Initiative. National Survey of Children with Special Health Care Needs: Interactive data search of the 2005/06 data. http://www cshcndata org/DataQuery/SurveyAreas aspx?yid=2 [serial online] 2006; Accessed March 3, 2011.

(12) Simon TD, Berry J, Feudtner C et al. Children with complex chronic conditions in inpatient hopsital settings in the United States. Pediatrics 2010;126:647-655.

(13) Golden SL, Nageswaran S. Caregiver voices: coordinating care for children with complex chronic conditions. Clin Pediatr 2012;51:723-729.

(14) Srivastava R, Stone BL, Murphy NA. Hospitalist care of the medically complex child. Pediatr Clin North Am 2005;52:1165-1187.

(15) Wise PH. The transformation of child health in the US. Health Aff (Millwood) 2004;23:9-25.

(16) Newacheck PW, Kim SE. A national profile of health care utilization and expenditures for children with special health care needs. Arch Pediatr Adolesc Med 2005;159:10-17.

(17) American Academy of Pediatrics, American Academy of Family Physicians and American College of Physicians-American Society of Internal Medicine. A consensus statement on health care transitions for young adults with special health care needs. Pediatrics 2002;110:1304-1306.

(18) American Academy of Pediatrics, American Academy of Family Physicians, American College of Physicians, Transitions Clinical Report Authoring Group. Supporting the health care transition from adolescence to adulthood in the medical home. Pediatrics 2011;128:182-200.

(19) Kelly AM, Kratz B, Bielski M, Rinehart PM. Implementing Transitions for Youth With Complex Chronic Conditions using the Medical Home Model. Pediatrics 2002;110:1322-1327.

(20) Oswald DP, Gilles DL, Cannaday MS, Wenzel DB, Willis JH, Bodurtha JN. Youth with special health care needs: transition to adult health services. Matern Child Health J 2013;17:1744-1752.

(21) Kasuki L, Marques NV, Nuez MJ, Leal VL, Chinen RN, Gadelha MR. Acromegalic patients lost to follow-up: a pilot study. Pituitary 2013;16:245-250.

(22) Delemer B, Chanson Ph, Foubert L et al. Patients lost to follow-up in acromegaly: results of the ACROSPECT study. European Journal of Endocrinology 2014;170:791-797.
(23) Attanasio R, Montini M, Valota M et al. An audit of treatment outcome in acromegalic patienst attending our center at Bergamo, Italy. Pituitary 2008;11:1-11.
(24) Lanoy E, Mary-Krause M, Tattevin P et al. Predictors identified for losses to follow-up among HIV-seropositive patients. Journal of Clinical Epidemiology 2006;59:829-835.
(25) Bowen ME, Henske JA, Potter A. Health Care Transition in Adolescents and Young Adults with Diabetes. Clinical Diabetes 2010;28:99-106.
(26) Goossens E, Stephani I, Hilderson D et al. Transfer of Adolescents With Congenital Heart Disease From Pediatric Cardiology to Adult Health Care. An Analysis of Transfer Destinations. Journal of the American College of Cardiology 2011;57:2368-2374.
(27) Blum RW, Garell D, Hodgman CH et al. Transition from child-centered to adult health-care systems for adolescents with chronic conditions. A position paper of the Society for Adolescent Medicine. J Adolesc Health 1993;14:570-576.
(28) Carroll G, Massarelli E, Opzoomer A, et al. Adolescents with chronic disease: are they receiving comprehensive health care? J Adolesc Health Care 1983;17:32-36.
(29) Schidlow D, Fiel S. Life beyong pediatrics. Transition of chronically ill adolescents from pediatric to adult health care systems. Med Cllin North Am 1990;74:1113-1120.
(30) Yeung E, Kay J, Roosevelt GE, Brandon M, Yetman AT. Lapse of care as a predictor for morbidity in adults with congenital heart disease. International Journal of Cardiology 2008;125:62-65.
(31) Frank M. Factors associated with non-compliance with a medical follow-up regimen after discharge from a pediatric diabetes clinic. Can J Diabetes Care 1996;20:13-20.
(32) Pacaud D, McConnell B, Huot C. Transition from pediatric to adult care for insuline-dependent diabetes patients. Can J Diabetes Care 1996;20:14-20.
(33) Pacaud D, Yale JF, Stephure D, Trussell R, Davies HD. Problems in transition from pediatric care to adult care for individuals with diabetes. Can J Diabetes Care 2005;29:13-18.
(34) Griffin SJ. Lost to follow-up: the problem of defaulters from diabetes clinics. Diabet Med 1998;15:S14-S24.
(35) Nakhla M, Daneman D, To T, Paradis G, Guttmann A. Transition to adult care for youths with diabetes mellitus: findings from a Universal Health Care System. Pediatrics 2009;124:e1134-e1141.
(36) Shamseer L, Moher D, Clarke M et al. Preferred reporting items for systematic review and meta-analysis protocols (PRISMA-P) 2015: elaboration and explanation. BMJ 2015;349.
(37) Gleeson H, Turner G. Transition to adult services. Arch Dis Child Educ Pract Ed 2012;97(3):86-92.
(38) Mackie AS, Rempel GR, Rankin KN, Nicolas D, Magill-Evans J. Risk factors for loss to follow-up among children and young adults with congenital heart disease. Cardiol Young 2012;22:307-315.
(39) Andemariam B, Owarish-Gross J, Grady J, Boruchov D, Thrall RS, Hagstrom JN. Identification of risk factors for an unsuccessful transition from pediatric to adult sickle cell disease care. Pediatr Blood Cancer 2014;61(4):697-701.
(40) Reid GJ, Irvine MJ, McCrindle BW et al. Prevalence and correlates of successful transfer from pediatric to adult health care among a cohort of young adults with complex congenital heart defects. Pediatrics 2004;113:e197-e205.
(41) Hazel E, Zhang X, Duffy CM, Campillo S. High rates of unsuccessful transfer to adult care among young adults with juvenile idiopathic arthritis. Pediatr Rheumatol Online J 2010;8:1-6.
(42) Wilson DB. Practical Meta-Analysis Effect Size Calculator. 2015. 30-1-2015.
(43) Slim K, Nini E, Forestier D, Kwiatiwoski F, Panis Y, Chipponi J. Methodological Index for Non-Randomized Studies (MINORS): Development and Validation of a New Instrument. ANZ Journal of Surgery 2003;73:712-716.

(44) Wojciechowski EA, Hurtig A, Dorn L. A natural history of adolescents and young adults with sickle cell disease as they transfer to adult care: a nedd for case management services. J Pediatr Nurs 2002;17:18-27.
(45) Mackie AS, Ionescu-Ittu R, Therrien J, Pilote L, Abrahamowicz M, Marelli AJ. Children and adults with congenital heart disease lost to follow-up: who and when? Circulation 2009;120:302-309.
(46) Gurvitz M, Valente AM, Borberg C et al. Prevalence and Predictors of Gaps in Care Among Adult Congenital Heart Disease Patients (The Health, Education and Access Research Trial: HEART-ACHD). Journal of the American College of Cardiology 2013;61:2180-2184.
(47) Foster E, Graham TP, Driscoll DJ et al. Task Force 2: Special health care needs of adults with congenital heart disease. Journal of the American College of Cardiology 2001;37:1176-1183.
(48) Christie D, Viner R. Adolescent development. BMJ 2005;330:301-306.
(49) Steinberg L. Cognitive and affective development in adolescence. Trends in cognitive sciences 2005;9:69-74.
(50) Ferlie EB, Shortell SM. Improving the quality of health care in the United Kingdom and the United States: A Framework for Change. The Milbank Quarterly 2001;79:281-315.

6 ADOLESCENTS' CONGENITAL HEART DISEASE-RELATED KNOWLEDGE AFTER TRANSFER TO ACHD CARE

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Abstract

Background: Adolescents with congenital heart disease (CHD) must take responsibility for their life and care. This requires that they have sufficient knowledge about their heart disease, treatment, and preventive measures. Thus, CHD-related education should be directed to adolescents. Research on adolescents' understanding and knowledge of CHD is limited.

Aims: What do adolescents with CHD know about their heart defect, treatment, and preventive measures necessary to avoid complications?

Methods: We addressed this question in a descriptive cross-sectional study of 91 adolescents with CHD (53% males; median age, 17 years). In this study, we assessed the subjects' knowledge of CHD using the Leuven Knowledge Questionnaire for Congenital Heart Disease.

Results: The results showed that the patients had adequate knowledge (>80% correct answers) about the need for regular follow-up, required diet, past treatment, and dental practices. They had moderate knowledge (50-80% correct answers*)* about the frequency of follow-up, occupational choices, medication regimen, and sexual activities. However, the patients had poor knowledge (<50% correct answers) of the name of their heart defect; the reasons for follow-up; the effects of competitive sports; the symptoms that reflect deterioration of their heart disease; the definition, characteristics, and risk factors of endocarditis; the possibility of recurrent episodes of endocarditis during their lifetime; the impact of smoking and alcohol on their heart disease; the hereditary nature of their condition; the suitability of intrauterine devices as contraceptives; the appropriateness of oral contraceptives; and the risks of pregnancy.

Conclusion: In conclusion, this study showed that the level of knowledge of adolescents with CHD has significant gaps.

Introduction

To date, several studies investigating the level of knowledge of patients with congenital heart disease (CHD) have been undertaken. Five studies have been conducted in children [1-5], and 9 studies have been performed in adults [6-14]. To our knowledge, only 4 studies have included adolescents among their subjects [3,4,5,15]. One study specifically targeted adolescent patients, focusing on their knowledge of bacterial endocarditis [15]. Other aspects of CHD, however, were not addressed. This means that information on the level of knowledge of adolescents with CHD is scant. Therefore, we designed a study to investigate what adolescents with CHD know about their heart defect, its treatment, and preventive measures necessary to avoid complications.

Methods

We recruited literate, Dutch-speaking adolescents with CHD to participate in our descriptive cross-sectional study. Adolescents were eligible for the study upon their initial visit to the Adult Congenital Heart Disease (ACHD) Program's clinic after their transfer from pediatric cardiology. At the University Hospitals of Leuven, Belgium, it is standard practice for a pediatric CHD patient to be transferred to adult-focused care when he/she reaches the age of 16 years, unless the patient is medically unstable [16]. Patients were excluded from our study if they had learning disabilities. In a 13-month period, 100 adolescents who met the inclusion criteria were asked to participate. One patient refused to participate due to lack of interest. Eight patients were excluded because of practical reasons. Hence, we recruited 91 adolescents with CHD; 53% were male and 47% female. Patients had a median age of 17 years. **Table 6.1** summarizes the demographic and clinical characteristics of the study sample.

Table 6.1: Demographic and clinical characteristics of 91 adolescents with CHD

Variable	n	(%)
Sex		
Male	48	(53)
Female	43	(47)
Median age (years)	17	(Q_1= 16; Q_3=18) (Range: 15-32 years)
Marital status		
Unmarried (living with parents)	87	(96)
Living together	2	(2)
Living alone	1	(1)
Married	1	(1)
Highest educational level		
Vocational high school	29	(31)
Technical high school	32	(34)
High school/College/University	30	(32)
Responsible for the daily management of care		
Parents	1	(1)
Patient	23	(25)
Patient and parents	67	(74)
Treatment		
Surgery	31	(34)
Medication	4	(4)
No treatment	30	(33)
Surgery and catheter intervention	7	(8)
Surgery, catheter intervention, and medication	2	(2)
Surgery and medication	6	(7)
Catheter intervention	8	(9)
Medication and catheter intervention	3	(3)
History of endocarditis	0	(0)
Number of pregnancies	0	(0)
Contraception (for female patients only)		
Pill	19	(43)
Other methods	1	(2)
No contraception	23	(55)
Primary medical diagnosis		
Ventricular septal defect	26	(29)
Coarctation of the aorta	14	(16)
Pulmonary valve stenosis	11	(12)
Transposition of the great arteries	5	(6)
Aortic valve stenosis	5	(6)
Tetralogy of Fallot	4	(5)
Atrial septal defect type secundum	3	(3)
Congenitally corrected transposition of the great arteries	3	(3)

Mitral valve regurgitation	3	(3)
Aortic valve regurgitation	3	(3)
Univentricular heart	2	(2)
Atrioventricular septal defect	2	(2)
Patent ductus arteriosus	2	(2)
Mixed aortic valve disease	1	(1)
Truncus arteriosus	1	(1)
Pulmonary atresia	1	(1)
Double aortic arch	1	(1)
Total anomalous pulmonary venous return	1	(1)
Marfan syndrome	1	(1)
Atrial septal defect type primum	1	(1)
Atrial septal defect type sinus venosus	1	(1)

Demographic and clinical variables were gathered during patient interviews and by means of patient medical records. The patients' CHD knowledge was assessed with the Leuven Knowledge Questionnaire for Congenital Heart Disease, which was developed by Moons and coworkers in 2001 [10]. We adapted the questionnaire based on our experiences in the first study. The most current version of the questionnaire consisted of 27 items and covered 5 domains, which were identified as relevant aspects of patients' knowledge about CHD: (1) knowledge on heart defect and treatment, (2) knowledge on prevention of complications, (3) knowledge on physical activities, (4) knowledge on sexuality and heredity, and (5) knowledge on contraception and pregnancy planning. The researchers evaluated each patient's answers as 'correct,' 'does not know,' or 'incorrect' or 'incomplete' [10].

When the patients arrived for their scheduled outpatient visit at the ACHD Program's clinic, a nurse from the advanced practice nursing team approached the patients and explained the aims and protocol of the study. After oral informed consent was obtained, the nurse asked the patient to fill out the knowledge questionnaire, as he/she sat in the waiting room. The nurse explicitly asked family members not to help the patient complete the questionnaire. Moreover, the patient was forbidden from consulting external sources. The advanced practice nurse checked the questionnaire for completeness and asked for additional information, if necessary. Upon completion, the patient and his/her parents entered the consultation room, where the advanced practice nurse used the completed questionnaire as a guide to provide appropriate patient education [17]. The Institutional Review Board (IRB) of the University Hospitals Leuven approved the study protocol.

The data were analyzed with SPSS 16.00. Descriptive statistics of demographic and clinical variables were expressed in percentages, medians, and quartiles. The knowledge variables were dichotomized as correct or incorrect answers (the latter included incomplete, does not know, and incorrect responses).

Results

Less than half of the patients knew the name of their heart defect, while only 28% could describe their heart defect or locate the lesion on a diagram (**Table 6.2**). The majority of the patients knew the frequency of follow-up and the need for regular follow-up, but only 46% indicated that the main purpose of follow-up was to detect clinical deterioration. Most of the patients had adequate knowledge about their past treatment. Of the 15 individuals who took medications on a regular basis, 53% knew the name of the medication. Almost all of the patients knew which diet they had to follow. A small number of the patients could identify symptoms that reflect deterioration of the heart disease: dizziness, shortness of breath, palpitations, chest pain, fainting, increasing fatigue, and swollen feet and legs (**Table 6.2**).

Only 21% of the adolescents could correctly define endocarditis, and only 1 adolescent recognized unexplained fever for > 5 days as the most characteristic sign of endocarditis (**Table 6.3**). The minority knew that endocarditis could recur, and 78% knew that they should not take antibiotics without consulting a doctor. Only a small number of patients knew the risk factors for endocarditis: contaminated needles, bacteria from skin infections, dental abscesses, poor nail and skin care, and body piercing and tattooing. The patients, however, had good knowledge of dental practices. The majority of patients incorrectly believed that smoking and alcohol consumption, respectively, were more harmful to them than to their healthy counterparts (**Table 6.3**).

Table 6.2: Frequency of patients' knowledge about the disease and its treatment (n=91)

Question	Correct	Incorrect	Does not know	Incomplete
1. What is the name of your heart defect?	41 (45%)	9 (10%)	33 (36%)	8 (9%)
2. Describe or indicate on the diagram where your heart is located.	25 (27%)	12 (13%)	46 (51%)	8 (9%)
3. How often do you have to come to the outpatient clinic for follow-up of your congenital heart disease?	71 (78%)	14 (15%)	6 (7%)	-
4. What is the main purpose of cardiac follow-up?	42 (46%)	0 (0%)	0 (0%)	49 (54%)
5. How has your heart condition been treated to date?	77 (85%)	2 (2%)	4 (4%)	8 (9%)
6. If you are on drug treatment, give the name, dose, schedule, reason or function, most important side effects, and interactions with other drugs or foods.*	8 (53%)	2 (13%)	1 (7%)	4 (27%)
7. If you experience side effects from your prescribed drugs, does this mean you should stop taking them?	37 (41%)	3 (3%)	50 (56%)	-
8. Do you have to follow a diet? If you answer yes, please indicate the type of diet.	86 (94%)	0 (0%)	5 (6%)	-
9. Mark all symptoms that may occur if your heart condition deteriorates and for which you have to contact your cardiologist.	8 (9%)	0 (0%)	39 (43%)	44 (48%)
10. If the congenital cardiologist informs you that everything is all right, does that mean that you don't need further follow-up?	79 (87%)	8 (9%)	4 (4%)	-

** Only 15 out of the 91 patients received drug treatments for their congenital heart disease at the time of the survey.*

Table 6.3: Frequency of patients' knowledge about preventive measures (n=91)

Question	Correct	Incorrect	Does not know
11. What is endocarditis?	19 (21%)	11 (12%)	61 (67%)
12. What is the most typical sign or symptom of endocarditis?	1 (1%)	19 (21%)	71 (78%)
13. Can you only get endocarditis once in your lifetime?	13 (14%)	3 (3%)	75 (83%)
14. Do you think the following factors contribute to the onset of endocarditis?			
Needle contamination (drug addicts)	18 (20%)	7 (8%)	66 (72%)
Smoking	7 (8%)	25 (28%)	59 (64%)
Bacteria from skin infections	10 (11%)	8 (9%)	73 (80%)
Dental abscesses	20 (22%)	8 (9%)	63 (69%)
Sexual activity	22 (24%)	1 (1%)	68 (75%)
Poor nail and skin care	7 (8%)	17 (19%)	67 (73%)
Body piercing and tattooing	14 (15%)	17 (19%)	60 (66%)
15. As you have a congenital heart disease, should you take antibiotics immediately (without consulting a physician) if you have fever?	71 (78%)	7 (8%)	13 (14%)
16. Should you have a dental check-up at least once a year?	76 (84%)	8 (9%)	7 (7%)
17. Should you take antibiotics before every visit to the dentist?	71 (78%)	18 (20%)	2 (2%)
18. Do bleeding gums need extra attention?	68 (75%)	10 (11%)	13 (14%)
19. Should you clean your teeth at least once a day?	87 (96%)	3 (3%)	1 (1.)
20. Is smoking more harmful for patients with congenital heart disease than for other people?	7 (8%)	64 (70%)	20 (22%)
21. Is consuming alcohol 3 times a day more harmful for patients with congenital heart disease than for other people?	22 (24%)	31 (34%)	38 (42%)

In this study, 39% of the adolescents knew that engaging in high-level competitive sports, requiring daily training, is not allowed (**Table 6.4**). A large proportion of the patients were aware that they had to choose an occupation that is not too physically demanding.

Table 6.4: Frequency of patient's knowledge about physical activity (n=91)

Question	Correct	Incorrect	Does not know
22. Are you allowed to take part in competitive sports that require daily training?	35 (39%)	42 (46%)	**14 (15%)**
23. Should you choose an occupation that is not too physically demanding, as you should be careful not to overexert yourself?	**68 (75%)**	**11 (12%)**	**12 (13%)**

About 78% of the patients knew that they were allowed to have sex if they felt capable of doing so (**Table 6.5**). Only 20% of the adolescents knew about the hereditary nature of their condition.

With regard to contraceptives, 14% and 35% of the female patients did not know whether intrauterine devices and oral contraceptives, respectively, were suitable or appropriate choices (**Table 6.5**). The majority of the female patients had insufficient knowledge of the risks of pregnancy.

Table 6.5: Frequency of patient's knowledge about reproductive issues (n=91)

Question	Correct	Incorrect	Does not know
24. In terms of sexual physical effort, can you do all what you feel you are able to do?	71 (78%)	1 (1%)	**19 (21%)**
25. What is the chance that your children will have congenital heart disease?	18 (20%)	24 (26%)	**49 (54%)**
26. Which contraceptives are the most advisable for you to use in light of your congenital heart disease? (only for women)			
Contraceptive pill	15 (35%)	-	**28 (65%)**
Intrauterine device	6 (14%)	3 (7%)	**34 (79%)**
27. Are you at risk for deterioration during pregnancy? (only for women)	**5 (12%)**	**11 (25%)**	**27 (63%)**

Discussion

As they grow older, adolescents with CHD presumably take responsibility for their own health and care. Transition programs should be implemented in order to prepare adolescents for this task [18-21]. A critical element of these transition programs is developmentally appropriate education for patients [18,22] that aim to improve the patients' level of knowledge and to increase awareness of adopting adequate health behaviors, while taking the transition through puberty into account. With the exception of one study that assessed bacterial endocarditis knowledge [15], studies that have specifically investigated the level of knowledge of adolescents with CHD, to the best of our knowledge, do not exist. Hence, we studied the level of knowledge in a sample of adolescents upon their transfer from pediatric cardiology to adult-focused care.

In general, our findings indicate that the level of knowledge of adolescents who recently transferred to adult care is poor. The findings of our study are to a certain extent comparable with those of prior investigations. In the studies of Veldtman [3] and Cetta [15], 30% to 69% of the patients were able to describe or provide the name of their heart defect. In the present study, 45% of the patients were within this range. On the other hand, there are also substantial differences between our study and previous ones. For instance, the level of knowledge about medication was considerably lower in our study (53%) than in that of Cetta [15]; and the knowledge of risk factors for endocarditis (8%-24%) was lower than that in Knirsch [5]. By contrast, our patients were more knowledgeable about other areas than patients of previous studies: definition of endocarditis (21% vs. 4%) [15]; preventive measures (75%-96% vs. 0%) [15]; and antibiotic prophylaxis (78% vs. 40%) [5,15]. Note, however, that the findings of the different studies are not comparable, because the age ranges, focus of the research, and measurements differed across the studies.

Currently, it is the policy of our institution to transfer adolescents from pediatric cardiology to the ACHD program when they reach the age of 16 years [16]. This policy is successful, with to date 84% of the patients have received specialist care after they have left pediatric cardiology [23]. A formal transition program, however, does not precede this transfer. The findings of the present study advocate such a transition program. Indeed, patient education is a critical element of transition programs [18,22], particularly because the responsibility of healthcare management shifts from the family to the patient [18]. To support adolescent patients to take responsibility for their healthcare, healthcare professionals should inform and instruct them before they transfer to adult-focused care. Although pediatric cardiologists and the patients' families already discuss several topics covered in the education program, this strategy apparently does not ensure that patients retain the information. Indeed, there is a huge disparity between the information provided, the information

understood, and the information retained. In addition, the growth rate of each adolescent varies; thus, their mental maturity, sense of responsibility, and self-care can differ widely. It is therefore of paramount importance to realize that the onset of puberty can hinder patients from being open to information and instruction by healthcare professionals. For this reason, ACHD programs should also continuously invest in patient education, to ensure that information is retained. Therefore, the integration of nurse specialists in ACHD teams is advocated [24-26].

Structured patient education has proved to be effective in increasing the level of knowledge in patients with CHD. At our ACHD program, the effects of a structured education program were evaluated using a pre-post design, with an interval of 5 years [27]. An improvement of >10% was observed for knowledge about the name of the heart defect, side effects of medication, symptoms of deterioration, risk factors for endocarditis, appropriate use of antibiotics, appropriateness of contraceptive pills and intrauterine devices, definition of endocarditis, and characteristics of endocarditis [27]. In another study, we demonstrated that advanced practice nursing teams can have an impact on the pregestational counseling of CHD patients [28]. Before the implementation of advanced practice nursing, 44% of female patients received cardiac follow-up during pregnancy. This proportion increased to 71% after the advanced practice nursing team implemented systematic educational interventions that contribute to a better understanding of the rationale for cardiac appointments during pregnancy and a better adherence to follow-up recommendations [28]. Admittedly, we do not have empirical evidence supporting the effectiveness of such an education program in adolescents. Further research in this respect is imperative. Furthermore, it is important to determine which aspects of knowledge are indispensable. This can help ACHD professionals to set priorities in the content of patient education programs, because too much information overloads patients and hinders information retention.

Some methodological limitations require that the results of this study be interpreted with caution. First, this study is a single center study conducted at an outpatient clinic. Therefore, we have to be careful in generalizing the study results. Second, the Leuven Knowledge Questionnaire for Congenital Heart Disease was developed in 2001 [10]. The scale was initially tested in 62 adults with CHD [10]. The content validity of the questionnaire was not examined in adolescents with CHD, neither was the ability of these adolescents to read and understand the questions in this instrument. Although we did not experience problems in this respect in the present study, further testing of the validity and reliability of the questionnaire in this specific population is needed. Third, the instrument is multidimensional, making the calculation of an overall knowledge score have little meaning. The absence of a total score may in some situations be an obstacle. It may be useful to find a way in which the level of knowledge of a patient can be aggregated into a single index value.

References

(1) Shebani SO, Miles HF, Simmons P, Stickley J, De Giovanni JV. Awareness of the risk of endocarditis associated with tattooing and body piercing among patients with congenital heart disease and paediatric cardiologists in the United Kingdom. Arch Dis Child 2007; 92:1013-1014.
(2) Beeri M, Haramati Z, Rein JJ, Nir A. Parental knowledge and views of pediatric congenital heart disease. Isr Med Assoc J. 2001;3:194-197.
(3) Veldtman GR, Matley SL, Kendall L, Quirk J, Gibbs JL, Parsons JM, Hewison J. Illness understanding in children and adolescents with heart disease. Heart 2000;84:395-397.
(4) Kendall L, Parsons JM, Sloper P, Lewin RJ. A simple screening method for determining knowledge of the appropriate levels of activity and risk behaviour in young people with congenital cardiac conditions. Cardiol Young 2007;17:151-157.
(5) Knirsch W, Hassberg D, Beyer A, Teufel T, Pees C, Uhlemann F, Lange PE. Knowledge, compliance and practice of antibiotic endocarditis prophylaxis of patients with congenital heart disease. Pediatr Cardiol 2003;24:344-349.
(6) Rogers P, Mansour D, Mattinson A, O'Sullivan JJ. A collaborative clinic between contraception and sexual health services and an adult congenital heart disease clinic. J Fam Plann Reprod Health Care 2007;33:17-21.
(7) Simko LC, McGinnis KA, Schembri J. Educational needs of adults with congenital heart disease. J Cardiovasc Nurs 2006;21:85-94.
(8) Chessa M, De Rosa G, Pardeo M, Negura GD, Butera G, Feslova V, Piazza L, Giamberti A, Bossone E, Carminati M. Illness understanding in adults with congenital heart disease. Ital Heart J. 2005;6:895-899.
(9) Dore A, de Guise P, Mercier LA. Transition of care to adult congenital heart centres: what do patients know about their heart condition? Can J Cardiol 2002;18:141-146.
(10) Moons P, De Volder E, Budts W, De Geest S, Elen J, Waeytens K, et al. What do adult patients with congenital heart disease know about their disease, treatment, and prevention of complications? A call for structured patient education. Heart 2001;86:74-80.
(11) Kantoch MJ, Collins-Nakai RL, Medwid S, Ungstad E, Taylor DA. Adult patients' knowledge about their congenital heart disease. Can J Cardiol 1997;13:641-645.
(12) Cetta F, Warnes CA. Adults with congenital heart disease: patient knowledge of endocarditis prophylaxis. Mayo Clin Proc 1995;70:50-54.
(13) Vogel M, Knirsch W, Lange PE. Schwere komplikationen durch nichtbeachtung der endokarditisprophylaxe wahrend zahnarztlicher eingriffe bei erwachsenen mit angeborenen herzfehlern. [Severe complications caused by inattention to endocarditis prevention during dental procedures in adults with congenital heart abnormalities]. Dtsch Med Wochenschr 2000;125:344–347.
(14) Cetta F, Graham LC, Lichtenberg RC, Warnes CA. Piercing and tattooing in patients with congenital heart disease: patient and physician perspective. J Adolesc Health 1999;24:160–162.
(15) Cetta F, Podlecki DC, Bell TJ. Adolescent knowledge of bacterial endocarditis prophylaxis. J Adolesc Health 1993;14:540-542.
(16) Moons P, Pinxten S, Dedroog D, Van Deyk K, Gewillig M, Hilderson D, et al. Expectations and experiences of adolescents with congenital heart disease on being transferred from pediatric cardiology to the adult congenital heart disease program. J Adolesc Health 2009; 44: 316-322.
(17) Moons P, De Geest S, Budts W. Comprehensive care for adults with congenital heart disease: Expanding roles for nurses. Eur J Cardiovasc Nurs 2002;1:23-28.
(18) Saidi A, Kovacs AH. Developing a Transition Program from Pediatric- to Adult-Focused Cardiology Care: Practical Considerations. Congenit Heart Dis 2009: 204-215.
(19) Berg SK, Hertz PG. Outpatient nursing clinic for congenital heart disease patients: Copenhagen Transition Program. J Cardiovasc Nurs 2007;22: 488–492.

(20) Hudsmith LE, Thorne SA. Transition of care from paediatric to adult services in cardiology. Arch Dis Child 2007;92:927–930.
(21) Moons P, Hilderson D, Van Deyk K. Implementation of transition programs can prevent another lost generation of patients with congenital heart disease. Eur J Cardiovasc Nurs 2008; 7: 259-263.
(22) Van Deyk K, Moons P, Gewillig M, Budts W. Educational and behavioral issues in transitioning from pediatric cardiology to adult-centered health care. Nurs Clin North Am 2004; 39: 755-768.
(23) Goossens E, Stephani I, Hilderson D, Gewillig M, Budts W, Van Deyk K, et al. Transfer destinations and risk factors for 'no follow-up' and 'no appropriate follow-up' in young adults with congenital heart disease after leaving pediatric cardiology. J Am Coll Cardiol 2010; 55: A42.E399.
(24) Moons P, Scholte op Reimer W, De Geest S, Fridlund B, Heikkila J, Jaarsma T, et al. Nurse specialists in adult congenital heart disease: The current state in Europe. Eur J Cardiovasc Nurs 2006; 5: 60-67.
(25) Moons P, Engelfriet P, Kaemmerer H, Meijboom FJ, Oechslin E, Mulder BJM. Delivery of care for adult patients with congenital heart disease: Results from the Euro Heart Survey. Eur Heart J 2006; 27:1324-1330.
(26) Moons P, Meijboom FJ, Baumgartner H, Trindade PT, Huyghe E, Kaemmerer H, on behalf of the ESC Working Group on Grown-up Congenital Heart Disease. Structure and activities of adult congenital heart disease programmes in Europe. Eur Heart J 2010; 31: 1305-1310.
(27) Van Deyk K, Moons P, Farroni N, Dedroog D, Pinxten S, Troost E, et al. Effects of a structured education programme for adult patients with congenital heart disease: a follow-up study. Acta Cardiol 2008;63:125-126.
(28) Moons P, Budts W, Costermans E, Huyghe E, Pieper PG, Drenthen W, on behalf of the ZAHARA investigators. Pregnancy-related health behavior of pregnant women with congenital heart disease: Room for behavioural change interventions. Congenit Heart Dis 2009; 4: 348-355.

7 PSYCHOMETRIC PROPERTIES OF THE HEALTH BEHAVIOR SCALE-CONGENITAL HEART DISEASE

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Abstract

Background: To optimize long-term outcomes, patients with congenital heart disease (CHD) should adopt health-promoting behaviors. Studies on health behavior in afflicted patients are scarce and comparability of study results is limited. To enlarge the body of evidence, we have developed the Health Behavior Scale-Congenital Heart Disease (HBS-CHD).

Aims: We examined the psychometric properties of the HBS-CHD by providing evidence for (a) the content validity; (b) validity based on the relationships with other variables; (c) reliability in terms of stability; and (d) responsiveness.

Methods: Ten experts rated the relevance of the HBS-CHD items. The item content validity index (I-CVI) and averaged scale content validity index (S-CVI/Ave); the modified multi-rater Kappa and proportion of missing values for each question were calculated. Relationships with other variables were evaluated using six hypotheses that were tested in 429 adolescents with CHD. Stability of the instrument was assessed using Heise's method; and responsiveness was tested by calculating the Guyatt's Responsiveness Index (GRI).

Results: Overall, 86.3% of the items had a good to excellent content validity; the S-CVI/Ave (0.81) and multi-rater Kappa (0.78) were adequate. The average proportion of missing values was low (1.2%). Because five out of six hypotheses were confirmed, evidence for the validity of the HBS-CHD based on relationships with other variables was provided. The stability of the instrument could not be confirmed based on our data. The GRI showed good to excellent capacity of the HBS-CHD to detect clinical changes in the health behavior over time.

Conclusion: We found that the HBS-CHD is a valid and responsive questionnaire to assess health behaviors in patients with CHD.

Introduction

As adolescents born with a congenital heart defect (CHD) transition to adulthood, they are supposed to increasingly take responsibility over their health. During this developmental transition, adolescents become more independent and search for their own identity, develop a social network of peers, and increasingly adhere to their own values, beliefs, and customs (1). This developmental stage is generally characterized by experimenting behaviors such as smoking tobacco, use of illicit drugs, and alcohol (2,3).

Although to date about 90% of children born with CHD reach adulthood (4), they remain susceptible for developing arrhythmias, ventricular dysfunction, endocarditis, and premature mortality (5,6). In order to prevent these complications and to optimize long-term outcomes, patients should conduct health-promoting behaviors. These behaviors comprise moderate use of alcohol, avoidance of smoking cigarettes, no use of illicit drugs, excellent oral hygiene, adequate engagement in physical activities, and good dietary habits (7).

Current literature shows that studies investigating health behaviors in adolescents and emerging adults with CHD are scarce. Studies demonstrated that rates of substance use among these youngsters were lower compared to general population samples or healthy peers (1,6,8,9). Nonetheless, 28% of adolescents and 54% of young adults with CHD performed significant substance use during the past 30 days (9). Excellent oral hygiene, characterized by annual dental visits, flossing, and daily brushing of teeth, was identified in a small proportion of patients (9,10). The comparability of these study results, however, is limited because no standardized method to collect data on health behavior exists to date (9). Although to date some self-administered questionnaires are available for assessing health behaviors in patients with CHD (1,9-12), to the best of our knowledge none of them cover all items relevant for afflicted patients comprehensively. Furthermore, previously developed surveys differ substantially in wording, components of health behavior measured, and time frames used to assess health risk behavior (1,9-12).

In order to enlarge the body of evidence and to support clinical practice in assessing health risk behaviors of patients with CHD, we developed the Health Behavior Scale-CHD (HBS-CHD). This scale is a comprehensive tool for measuring and detecting potentially health-compromising behaviors in patients with CHD. Hence, the HBS-CHD contains items that are particularly relevant for afflicted patients, because these components may worsen patients' outcomes.

However, to use this tool in research or clinical practice, psychometric properties should be determined. The aim of this study was therefore to assess the psychometric properties of the HBS-CHD.

Methods

Development of the HBS-CHD

The HBS-CHD (**Appendix Figure 7.3**) was partially based on existing instruments comprising questions regarding health behavior in adolescents or adults (9,13-21). Twenty-five questions on 22 components of health risk behavior in individuals with CHD were formulated. Four questions regarding consumption of alcohol were based on the Alcohol Use Disorders Identification Test (AUDIT) (13,19,20). Three questions on the use of tobacco during the past month were based on the Youth Risk Behavior Survey (YRBS) (15,16,18). Seven questions, developed for a population-based study in the Netherlands, asked respondents about the use of (illicit) drugs, sleeping pills, sedatives, and tranquillizers during the past month (21). Information on dental hygiene was obtained through four questions derived from the Self-reported Health Risk Behaviors questionnaire (9,22). Seven questions pertained to physical activity levels, which were inspired by the Baecke questionnaire for the Measurement of a Person's Habitual Physical Activity (14,17). Levels of physical activity were operationalized using the classification schemes published by Godin et al. (23) and Durnin et al. (24). These questions were put in a specific lay-out to guide respondents through the HBS-CHD instrument.

Psychometric properties

To evaluate the psychometric properties of the HBS-CHD, we used the approach described and terminology used in the Standards for Educational and Psychological Testing (25). More specifically, we evaluated the evidence for content validity; validity evidence based on relationships with other variables; reliability evidence based on stability; and responsiveness evidence. These evaluations were undertaken concomitantly.

Content validity of the HBS-CHD

To evaluate the content validity, ten experts (three nurses, seven cardiologists) in pediatric cardiology and Adult Congenital Heart Disease were invited to rate the relevance of all 22 HBS-CHD items, using a 4-point rating scale (*1=not relevant; 4=very relevant*) (26). Free text space was provided to give additional comments. Calculation of both item content validity index (I-CVI) and averaged scale level content validity index (S-CVI/Ave) was performed. The excellence of the content

validity was assessed using generally accepted cut-off values (≥0.78 I-CVI for ≥6 experts; ≥0.80 S-CVI/Ave) (26-29). To adjust for agreement by chance, the modified multi-rater Kappa (κ*) was calculated. Cut-off values for κ* were <0.40 for poor, ≥0.40 and <0.60 for fair, ≥0.60 and <0.75 for good, and ≥0.75 for excellent item relevance (30,31).

Furthermore, the proportion of missing values for each item of the HBS-CHD was determined. This is a parameter of how intelligible an item is (25).

Validity evidence based on relationships with other variables

Evidence based on relationships with other variables was evaluated by testing six hypotheses (1,8-12). Population-based health behaviors surveillance systems (18,32) and a study on health behaviors in individuals with CHD (9) showed that risky health behaviors are more prevalent in (emerging) adults than in adolescents. Even within the groups of adolescents, increasing trends of health risk behaviors were observed (32). This brought us to formulate the following three hypotheses:

Hypothesis 1: The prevalence of substance use in adolescents and emerging adults with CHD is positively associated with increasing age (9,32).

Hypothesis 2: The prevalence of preventive dental hygiene in adolescents and emerging adults with CHD is negatively associated with increasing age (9).

Hypothesis 3: The prevalence of overall health-risk behaviors is positively associated with increasing age (18,32).

Studies that compared health behaviors of patients with CHD and healthy controls showed better behaviors in patients with regard to the use of alcohol, illicit drugs, and tobacco, but worse behaviors in terms of dental practices (1,8-10,12). Therefore, we formulated two hypotheses on the difference between patients and healthy controls.

Hypothesis 4: The prevalence of substance use in adolescents and emerging adults with CHD is lower than that of controls from the general population (1,8-10,12).

Hypothesis 5: The prevalence of preventive dental hygiene in adolescents and emerging adults with CHD is lower than that of controls from the general population (9,10).

Finally, we formulated a hypothesis regarding the relation between the Baecke's sport score (14) and the HBS-CHD physical exercise score. Although there is no gold standard in the self-report of physical activities, the Baecke questionnaire is well validated (33,34). A good relationship between the Baecke sports score and the HBS-CHD physical exercise score, which calculation was based on the Baecke's algorithm, would support the validity evidence.

Hypothesis 6: There is a high correlation (≥0.70) between the physical exercise score of the HBS-CHD and the Baecke's Sport Score (14).

If the hypotheses are confirmed by empirical testing, the validity of the instrument under study is supported (25,35).

Reliability evidence based on stability

For the evaluation of the stability of the HBS-CHD, a traditional test-retest is not applicable because behaviors are not stable in itself. Therefore, an alternative approach that is able to distinguish the stability of the concept (i.e., health behavior) from the stability of the tool (i.e., HBS-CHD) is used. We employed the technique as described by Heise (1969) (36,37), which requires four measurement points.

Responsiveness evidence

Responsiveness is "the ability of an instrument to record meaningful or clinically relevant changes in the patient's clinical state (e.g., health behavior) over time" (38). We assessed the internal responsiveness of the HBS-CHD, defined as " the ability of a measure to change over a predefined time frame" (39). Internal responsiveness can be evaluated with the use of a repeated measures design evaluating the changes in scale scores in a single sample of patients (39). More specifically, we calculated Guyatt's Responsiveness Index (GRI) (40).

Study population for the hypothesis-testing, assessment of reliability and responsiveness

We examined the validity in relation with other variables, reliability and responsiveness of the HBS-CHD as part of a four-wave longitudinal project, spanning three years (at nine-month intervals): the i-DETACH project (**I**nformation technology **D**evices and **E**ducation program for **T**ransitioning of **A**dolescents with **C**ongenital **H**eart disease). Eligible patients were selected from the database of pediatric and congenital cardiology of the University Hospitals Leuven, Belgium. Patients were included if: they had a confirmed diagnosis of CHD, defined as structural abnormalities of the heart and/or great intrathoracic vessels that are actually or potentially of functional significance (41); aged

14-18 years at the start of the study on October 22, 2009; last cardiac outpatient visit at our tertiary care center performed ≤ 5 years ago; being able to read and write Dutch; and the availability of valid contact details. Patients were excluded if they had cognitive and/or physical limitations that inhibit the patient to fill out questionnaires; if the patient previously underwent heart transplantation; and if patients and/or their parents did not consent to participate.

Overall, 498 patients met these criteria. A total of 429 adolescents (86%) participated in the first wave of the study; 398 patients (80%) partook in the second wave; and 363 patients (73%) completed the questionnaires in the third wave. In all, 348 participated in wave 1, 2, and 3. Wave 4 is currently still in progress. In June 2012, a total of 231 respondents had participated in the four subsequent waves.

At Wave 1, control subjects, comprising peers from the general population, were recruited at four secondary schools in two regions of Belgium. Matching (1:1) was performed based on gender and age, resulting in 401 patients matched with a control subject (93.5%).

Measurements and procedure

Data were obtained using the HBS-CHD and a modified version of the Baecke questionnaire. The Baecke questionnaire is a self-report instrument assessing the habitual physical activity of adults (14), which has been extensively used during the past two decades in physical activity research (42;43). Although no gold standard for the self-report of physical activity levels exists, the Baecke questionnaire was found to be a standard of reference that was validated against the double labeled water technique and a tri-axial accelerometer (33,34). The Baecke questionnaire comprises three dimensions: (a) physical activity at work; (b) sports activity during leisure time; and (c) physical, non-sports activity during leisure time. Since our study respondents are all school-attending adolescents, we used a modified version of the Baecke questionnaire that collects data on leisure time and sport physical activity indices. For the purpose of the present study, we only used the Baecke Sport Score.

Each wave, all eligible adolescents with CHD received a package by surface mail, which included a set of questionnaires, an information letter, an informed consent form (for parents and adolescents), and a pre-stamped and addressed return envelope. To obtain a high response rate, a modified Dillman's approach was used (44). More detailed information on this approach can be found in a related article (45). The study was approved by the Institutional Review Board of the

University Hospitals Leuven and the investigation was conducted in keeping with the principles outlined in the Declaration of Helsinki (46).

Statistical analysis

To test the six hypotheses, we first calculated summary scores. A 'physical exercise score' was calculated based on the usual time (in hours) spent per week in various types of physical exercise, including the walk or bike ride to school or work (there and back), multiplied by the average energy expenditure per unit of time (MJ/h), as derived from Baecke (14). This physical exercise score ranges from 0 to ∞, with higher scores indicating higher levels of physical exercise. Furthermore, the Baecke's Sport Score was calculated by multiplying the intensity of the practiced sport, the amount of time weekly playing that sport and the proportion of the year in which the sport was practiced (14).

A 'substance use score', ranging from 0 to 3, was calculated based on the presence of (a) binge drinking at least monthly, (b) use of ≥1 of 7 predefined drugs once a month or less, and (c) smoking of cigarettes. A 'dental hygiene risk score', varying between 0 and 3, was calculated based on the reporting that (a) the patient did not visit the dentist annually, (b) did not daily brush, and (c) did not floss his teeth. Finally, an 'overall health risk score' was computed based on the individuals' substance use score, dental hygiene risk score, and the absence of sport participation. This latter score ranges from 0 to 7. These latter three risk scores are recoded to a scale ranging from 0 (*no risk*) to 100 (*maximum risk*). In other words, a higher risk score represents a worse health behavior.

Statistical analyses were performed using SPSS version 17.0 (SPSS Inc., Chicago, Illinois). Descriptive statistics of the three risk scores were expressed in terms of means and standard deviations for reasons of clarity and comparability. Differences in median risk scores between different age cohorts were tested using the non-parametric Jonckheere-Terpstra trend test. Intra-individual changes in median subscale scores over a period of 18 months were evaluated using the Friedman's test. Differences between patients and matched controls were tested using the McNemar test for nominal data and the Wilcoxon-signed rank test for ordinal data. To test the convergent validity of the HBS-CHD compared with the Baecke's questionnaire, we investigated the relationship between the HBS-CHD physical exercise score and the Baecke's Sport Score, both measured on a continuous scale, by calculating the Pearson's product moment correlation coefficient.

To test the stability of the HBS-CHD, we used the algorithms described by Heise (37) employing data of 4 measurement points. We calculated reliability and stability coefficients for the

three risk scores: substance use risk score; dental hygiene risk score; and overall health risk score. The reliability coefficient of the instrument was calculated based on the equation:

$$r_{xx} = (r_{12} \times r_{23})/r_{13}$$

where the *r*s are the test-retest correlations (37). To test the assumptions underlying this technique, the product of r_{14} and r_{23} must be very close to the product of r_{13} and r_{24} (37).

To evaluate the internal responsiveness of the HBS-CHD, we calculated the Guyatt's Responsiveness Index (GRI). This GRI is the ratio of the minimally clinically important difference (i.e., a priori determined delta) divided by the root square of two times the Mean Squared Error of the ANOVA for repeated measures (39,40). We determined that a clinically significant change in health behavior is represented by one additional behavioral risk factor, reflecting a delta-value of 1. Cut-off levels for the interpretation of the GRI are 0.20 for poor; 0.50 for moderate; 0.80 for good; and >1 for excellent responsiveness (40).

Results

Sample characteristics

Adolescents with CHD in our sample had a median age of 16.3 years (Q1=15.3y; Q3=17.3y) (**Table 7.1**). The most common diagnosed heart defect was a Ventricular Septal Defect (18.2%), followed by aortic valve abnormality (16.1%) and secundum Atrial Septal Defect (13.1%). The majority of respondents had a moderately complex heart defect (47.6%), whereas mild and complex heart lesions were diagnosed in 40.6% and 11.9%, respectively. Additional details on sample characteristics can be found in **Table 7.1**. Participants did not differ on sex (χ^2=0.163;p>0.05) and age (U=593.0;p>0.05) from non-participants. However, differences were found on complexity of CHD (F=9.255;p<0.05), with the group of non-responders having relatively more mild and fewer moderate congenital heart lesions.

Content validity

Fourteen out of 22 (63.6%) HBS-CHD scale items were rated with an excellent content validity (I-CVI ≥0.78; κ^*≥0.75), and five other items (22.7%) were evaluated with a good content validity (κ^*≥0.60-<0.75). Two items ('11. Use of hallucinogenic mushrooms during last 12 months?' and '20. If yes, how long does it take by bike or on foot (there and back)?'*)* obtained an I-CVI of 0.60 and a κ^* of 0.50, representing a fair content validity (κ^*≥0.40-<0.60). One item (*'18. How often do you floss your teeth?')* was evaluated as having a poor content validity (I-CVI=0.50; κ^*=0.34). The S-CVI/Ave was 0.81 and the overall instrument's Kappa was 0.78, which reflects an adequate content validity

(26;29). If the three items with poor or fair content validity would be removed from the scale, the S-CVI/Ave would increase to 0.85. However, because of clinical and theoretical considerations, these items were kept in the questionnaire to allow further analysis of the psychometric properties. The overall proportion of missing values over all scale items was low (1.2%). At item level, the proportion of missing values ranged from 0.0% to 5.3%. Questions regarding the frequency of flossing (5.3%) and brushing the teeth (3.9%) yielded the highest rate of missing values (**Table 7.2**).

Validity based on relationships with other variables

A comparison of the risk scores for substance use, dental hygiene and overall health risk, according to the age group, was performed (**Table 7.3**). In order to test the intra-individual evolution in risk scores over a period of 18 months, we compared these risk scores in adolescents with CHD (n=348) compared across wave 1, 2, and 3 (**Table 7.4**).

Data revealed that there is an increasing trend in substance use when adolescents with CHD are growing older (Z = 3.71; p<0.001) (**Table 7.3**). During an 18-month interval, a significant intra-individual increase in substance use was found (χ^2=38.138; p<0.001) (**Table 7.4**). Hence, the first hypothesis can be confirmed.

The dental hygiene risk score increased in patients with CHD aged ≤16.9 years, but declined afterwards. Trend analysis showed no statistically significant evolution (Z= -1.32; p=0.19) (**Table 7.3**). Intra-individual comparisons confirmed that the dental hygiene risk score remained relatively stable over an 18-month period (χ^2=0.258; p=0.879) (**Table 7.4**). These results did not confirm our second hypothesis.

Analysis of the overall health risk scores demonstrated increased scores in patients until the age of 16.9 years, although this trend was not statistically significant (Z=1.37; p=0.17) (**Table 7.3**). Intra-individual analysis, however, showed a significant increase in the overall health risk of patients with CHD (χ^2=14.983; p=0.001). Thus, our third hypothesis could be supported.

Table 7.1: Demographic and clinical characteristics of adolescents with congenital heart disease (n=429)

Variables		
Sex n (%)		
	Male	229 (53.4)
	Female	200 (46.6)
Age (median; Q1-Q3)		16.3y;15.3-17.3y
Primary diagnosis of CHD n (%)		
	Hypoplastic left-heart syndrome	2 (0.5)
	Univentricular physiology	4 (0.9)
	Tetralogy of Fallot	11 (2.6)
	Double-outlet right ventricle	12 (2.8)
	Double-inlet left ventricle	1 (0.2)
	Truncus arteriosus	1 (0.2)
	Transposition of the great arteries (TGA)	26 (6.1)
	Congenitally-corrected TGA	5 (1.2)
	Coarctation of the aorta	43 (10.0)
	Atrioventricular septal defect	6 (1.4)
	Atrial septal defect, type 1	4 (0.9)
	Ebstein malformation	2 (0.5)
	Pulmonary valve abnormality	38 (8.9)
	Aortic valve abnormality	69 (16.1)
	Aortic abnormality	9 (2.1)
	Left ventricle outflow tract obstruction	5 (1.2)
	Atrial septal defect, type 2	56 (13.1)
	Ventricular septal defect	78 (18.2)
	Mitral valve abnormality	37 (8.6)
	Pulmonary vein abnormality	9 (2.1)
	Other	11 (2.6)
Complexity of primary CHD diagnosis [60] n (%)		
	Complex	51 (11.9)
	Moderate	204 (47.6)
	Simple	174 (40.6)
Cardiac surgery for CHD n (%)		
	Yes, at least one cardiac surgical intervention	200 (53.4)
	No	229 (46.6)
Current level of education n (%)		
	High school/College/University	194 (47.0)
	Vocational high school	128 (31.0)
	Technical high school	84 (20.3)
	Education for adolescents with special needs	7 (1.7)

Table 7.2: Content validity: Analysis of item content validity index (I-CVI), modified multi-rater Kappa (κ*) and missing values

Item of the HBS-CHD	I-CVI	K*	Evaluation of K*[30]	Missing values n(%)
1. Do you consume alcohol from time to time?	1.00	1.00	Excellent	0 (0.0)
1a. If yes, how often?	1.00	1.00	Excellent	0 (0.0)
2. When consuming alcohol, how many glasses do you have on average?	0.80	0.79	Excellent	0 (0.0)
3. How often do you drink 6 glasses or more on one occasion?	0.70	0.66	Good	3 (1.1)
4. Do you smoke cigarettes occasioanlly or regularly?	1.00	1.00	Excellent	1 (0.3)
5. During the last 30 days, on how many days did you smoke cigarettes?	0.70	0.66	Good	0 (0.0)
6. During the last 30 days, on the days you smoked, how many cigarettes did you smoke a day?	1.00	1.00	Excellent	0 (0.0)
7. How often, in the last 12 months, did you take the following drugs?	-	-	-	-
7a. Cannabis (marihuana, hash)	0.80	0.77	Excellent	7(1.9)
7b. XTC (ectasy)	0.80	0.79	Excellent	10 (2.8)
7c. Cocaine	0.80	0.79	Excellent	10 (2.8)
7d. Hallucinogenic mushrooms	0.60	0.50	Fair	10 (2.8)
7e. Speed	0.80	0.79	Excellent	10 (2.8)
7f. Sleeping pills, sedatives, or tranquilizers	0.70	0.66	Good	10 (2.8)
7g. Other (illicit) drugs	0.70	0.66	Good	10 (2.8)
8. Have you been to the dentist in the past year?	1.00	1.00	Excellent	0 (0.0)
9. If not, when did you last go to the dentist?	0.90	0.90	Excellent	0 (0.0)
10. How often do you brush your teeth?	0.90	0.90	Excellent	14 (3.9)
11. How often do you floss your teeth?	0.50	0.34	Poor	19 (5.3)

HBS-CHD = Health Behavior Scale – Congenital Heart Disease; I-CVI = Item Content Validity Index; K = Modified multi-rater Kappa*

Item of the HBS-CHD	I-CVI	K*	Evaluation of K*[30]	Missing values n(%)
12. Do you regularly walk or cycle to school or to your place of work?	0.70	0.66	Good	0 (0.0)
13. If yes, how long does it take by bike or on foot (there and back)?	0.60	0.50	Fair	1 (0.4)
14. Do you regularly practice a sport (this includes school sport but not the bike ride or walk to school or to your workplace)?	1.00	1.00	Excellent	1 (0.3)
15. During a 7-day week, how many hours of the following physcial activities do you do?	0.90	0.90	Excellent	-
15a. Sports or activities that are very physically demanding, which increase your pulse (e.g., football, basketball, ...)	-	-	-	0 (0.0)
15b. Sports or activities that are moderately physically demanding and where, afterwards you don't feel exhausted or worn out (e.g., jogging, ballet, ...)	-	-	-	0 (0.0)
15c. Sports or activities with minimal physical effort or gentle exertions (e.g., golf, yoga, ...)	-	-	-	0 (0.0)
15d. Sport at school	-	-	-	0 (0.0)

HBS-CHD = Health Behavior Scale – Congenital Heart Disease; I-CVI = Item Content Validity Index; K = Modified multi-rater Kappa*

Comparison of the prevalence of binge drinking (i.e., ≥6 glasses of alcohol during one occasion) between adolescents with CHD and matched controls from general population, showed that significantly more controls performed binge drinking than patients with CHD ($p<0.001$). Furthermore, smoking of cigarettes and use of drugs during the past 12 months was significantly less prevalent in adolescents with CHD than peers ($p<0.001$). This corresponds with a significantly lower substance use score in patients than in matched controls ($Z=-6.38$; $p<0.001$). This corroborates the fourth hypothesis.

With regard to dental hygiene, more adolescents with CHD have an annual visit with their dentist than matched peers, although fewer patients report daily brushing of teeth or flossing of teeth. For the difference in flossing, statistical significance was reached ($p<0.001$). Furthermore, the dental hygiene risk score was significantly higher in patients with CHD compared to controls ($Z=-2.05$; $p=0.04$). Thus, the fifth hypothesis can be confirmed.

Finally, analysis showed the HBS-CHD physical exercise score to be significantly correlated with the Baecke's Sport Score (Pearson's $r=0.75$; $p<0.001$). This means that 56% of the variance (R^2) in the HBS-CHD physical exercise score could be explained by the Baecke's Sport Score (**Figure 7.2**). Therefore, our sixth hypothesis was confirmed.

Figure 7.2: Scatterplot of Health Behavior Scale – Congenital Heart Disease (HBS-CHD) physical exercise score versus Baecke's Sport Score

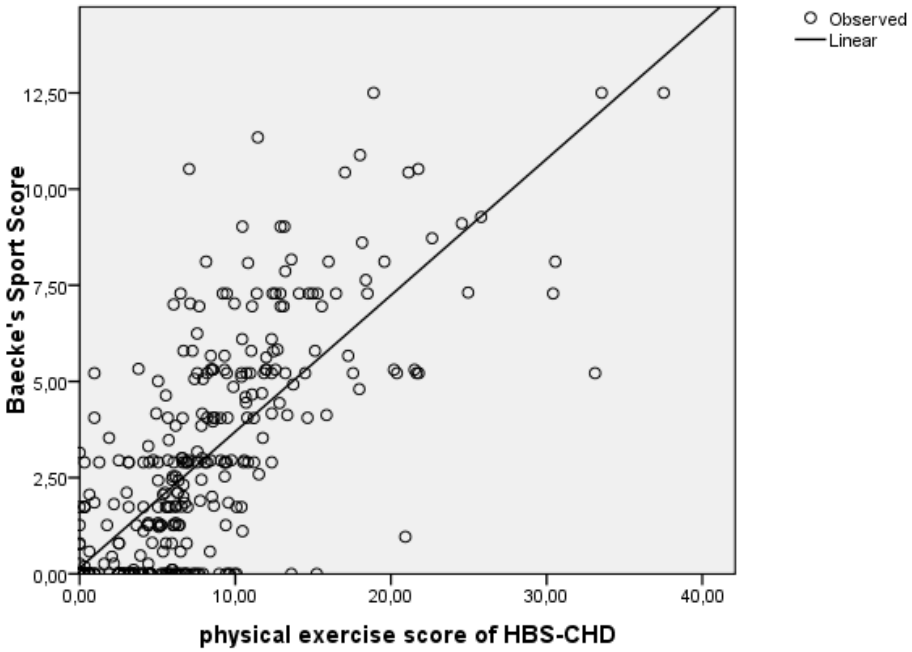


Table 7.3: Substance use, dental hygiene and health risk scores in adolescents with CHD according to their age group (n=424)

Risk score	14-14.9 years (n=70)	15-15.9 years (n=105)	16-16.9 years (n=112)	17-18.9 years (n=137)	Test statistics
Substance use risk score ($\bar{x}$±SD)	0.96 ± 8.00	5.70 ± 18.76	8.32 ± 20.28	8.72 ± 19.06	Z = 3.71; p<0.001
Dental hygiene risk score ($\bar{x}$±SD)	26.53 ± 23.14	26.49 ± 19.27	31.71 ± 25.52	22.73 ± 20.88	Z= -1.32; p=0.19
Total health risk score ($\bar{x}$±SD)	14.84 ± 12.85	16.56 ± 13.42	19.92 ± 15.96	17.62 ± 14.22	Z=1.37;p=0.17

All risk scores are transformed to a scale ranging from 0 to 100.
SD = standard deviation; Z = Z-score for Jonckheere-Terpstra trend test.

Table 7.4: Comparison of substance use, dental hygiene and health risk scores in adolescents with congenital heart disease (CHD), 18 month interval (n=348)

Risk score	Wave 1	Wave 2	Wave 3	Test statistics
Substance use risk score ($\bar{x}$±SD)	5.41 ± 16.39	7.91 ± 18.71	11.10 ± 22.65	χ^2 = 38.138; p<0.001
Dental hygiene risk score ($\bar{x}$±SD)	26.27 ± 22.32	26.08 ± 21.83	25.42 ± 23.24	χ^2 = 0.258; p=0.879
Health risk score ($\bar{x}$±SD)	16.65 ± 13.66	18.94 ± 14.88	19.90 ± 17.04	χ^2 = 14.953; p=0.001

All risk scores are transformed to a scale ranging from 0 to 100.
SD = Standard deviation

Reliability based on stability

Using Heise's method (37), we found a reliability coefficient of 1.08 for the substance use risk score; 0.37 for the dental hygiene risk score; and 0.57 for the overall health risk score. The underlying assumptions for this technique were violated for data on the substance use risk score, resulting in a coefficient >1. For the other two risk scores, the assumptions were fulfilled. The reliability coefficients were moderate to low. Based on these findings, the stability of the HBS-CHD over 9 month intervals could not be confirmed in the present study.

Responsiveness

The GRI was found to be 1.58 for the substance use risk score; 1.38 for the dental hygiene risk score; and 0.95 for the overall health risk score. This means that the HBS-CHD has a good to excellent capacity in detecting clinical changes in health behavior of patients with CHD over time.

Discussion

Although the importance of health-promoting and preventive behaviors in patients with CHD is well-established, a valid and comprehensive instrument to assess these behaviors is currently lacking. Therefore, we developed the comprehensive Health Behavior Scale-CHD scale (HBS-CHD), which relied in part on four existing questionnaires (9,13-15,17-20). The use of the HBS-CHD allows to calculating four summary risk scores: physical exercise score; substance use risk score; dental hygiene risk score; and total health risk score. In order to use this scale in research and clinical practice, we evaluated some psychometric properties of the instrument.

Our study revealed that 19 of the 22 items (86.3%) of this scale had a good to excellent content validity. The overall scale content validity was found to be adequate since S-CVI/Ave was 0.81 (26,29). Two items received an I-CVI <0.60 and ≥0.40 which corresponds to a fair content validity. Four experts commented that the use of hallucinogenic mushrooms as a drug is rare in the Belgian population. Therefore, these experts rated this item as irrelevant. However, because we aimed at developing a comprehensive health behavior questionnaire that is also applicable in an international context, we wanted to keep this question in our scale. Indeed, the use of hallucinogenic mushrooms is more prevalent in other countries than in Belgium (47). The second question, of which the relevance was found to be fair, concerned the duration of the bike ride or walk from home to school or work. Five experts rated this question as being not relevant but, unfortunately, gave no additional comments or suggestions. However, to be able to determine whether patients perform physical activities in accordance with general guidelines (48), we prefer to keep this question in the HBS-CHD.

The relevance of one item was assessed to be poor: the frequency of flossing the teeth. The importance of excellent dental care in patients with CHD is well established. Several guidelines recommend annual visits to the dentist, daily brushing of teeth, and the administration of antibiotics prior to specific dental procedures as essential components to prevent infective endocarditis (IE) (49,50). The relationship between flossing teeth and IE is controversial (49-56). On one hand, it is known that flossing may increase the occurrence of transient bacteremia, and thus may amplify the risk for IE (57). On the other hand, flossing teeth is an essential element of good dental hygiene, which in its turn can avoid IE. In order to prevent the formation of caries, patients should brush their teeth daily and floss their teeth at least weekly (55,58). Since the benefits of good dental hygiene, which includes interdental flossing, outweigh the risk for IE due to bacteremia, we keep this item in the HBS-CHD. Although none of the panel experts suggested adding additional items to the HBS-CHD,

one could argue that our scale should also cover aspects of healthy eating and weight control as these are potential risk factors for the development of cardiovascular disease in cardiac patients.

Furthermore, analysis of the missing values showed that the average proportion of missing values was low. Only for the questions regarding the frequency of flossing and brushing the teeth, the proportion of missing values was somewhat higher. Hence, there is sufficient evidence to consider the content of the HBS-CHD as valid.

Validity evidence based on relationships with other variables was tested with six hypotheses. The first three hypotheses pertained to substance use, dental hygiene, and overall health risk behaviors, and their relationship with age. We analyzed differences in risk scores across four age groups, and investigated the intra-individual evolution in patients over an 18-month period. These results provided evidence for hypotheses 1 and 3. We did not find support to confirm hypothesis 2, regarding dental hygiene. Based on the comparison of the prevalence of binge drinking, smoking, use of drugs, and annual dental visits, the fourth and fifth hypotheses stating that substance use and preventive dental hygiene measures are less prevalent in adolescents with CHD compared to peers, was confirmed. Finally, our last hypothesis on the relationship between the HBS-CHD physical exercise score and the Baecke's Sport Score provided evidence for the convergent validity of the HBS-CHD. Since five out of six proposed hypotheses were confirmed, the validity of the HBS-CHD based on relationships with other variables was generally supported.

Analysis of the reliability coefficients using the method of Heise (37) revealed that we could not confirm the stability of the HBS-CHD over a nine-month period of time. We assume that the nine-month intervals that we used in our study design were not optimal in order to assess the stability of our scale. Hence, further research on the instrument's stability is needed, in which shorter intervals between the measurements are required. On the other hand, the responsiveness of this scale could be supported.

Methodological limitations

This study aimed to assess some psychometric properties of the HBS-CHD. We provided evidence to support the content validity and evidence on relationships with other variables of this scale. Other aspects of validity, such as validity on response processes; validity on internal structure; and predictive validity with respect to consequences, were not investigated. Assessing the validity based on response processes necessitates specific research designs. Indeed, participant's response

processes could be evaluated using cognitive interviewing or observations during questionnaire completion (25). The validity on the internal structure is traditionally investigated using factor analysis (25,35). Several arguments were found against the use of exploratory factor analysis on the HBS-CHD scale. First, scale items are measured using several scale levels (e.g. nominal and ordinal data). Second, our scale comprises items aiming to screen patients for the use of alcohol, tobacco, etc. The use of these dichotomous items results in a large number of missing values for the sub-items when an item was not applicable to the patient. Factor analysis can only be performed on a dataset without missing values. Third, health behaviors are not necessarily interrelated (e.g. a patient who use alcohol does not necessarily use illicit drugs or smokes cigarettes), and a high frequency of alcohol consumption does not necessarily mean that the person drinks a high volume per occasion. Fourth, the analysis of a correlation matrix revealed that some items of our scale do not correlate with any other item; that a large number of items had a correlation coefficient <0.30; and that negative coefficients were observed. For all these reasons, the performance of exploratory factor analysis is not appropriate and not permitted on the HBS-CHD.

Assessment of the validity on the intended or unintended consequences (25) has limited relevance for validity testing of the HBS-CHD, because its relevance lies more in educational and employment testing than in testing clinical phenomena.

For reliability, we evaluated the instrument's stability. Other aspects of reliability, such as interrater reliability and internal consistency (25), were not tested. Since the HBS-CHD is a self-administered questionnaire, interrater reliability is not relevant. The same is true for the internal consistency. The items of the HBS-CHD are not supposed to measure one common concept. In addition, a Cronbach's alpha assumes that the items of the scale are correlated with each other at a level of 0.30 or above, because they are supposed to measure a common entity (59). In order to check this assumption, a correlation matrix was constructed to examine the direction and magnitude of correlations between the items of the instrument. We found that a number of items did not correlate to any other item, and that negative correlations were found. Hence, the calculation of Cronbach's alpha is not appropriate and not permitted.

Conclusion

The HBS-CHD was developed as a brief questionnaire to assess the health risk behaviors of adolescents, emerging adults and adults with CHD. The present study provided evidence for the content validity and on relationships with other variables, and on the responsiveness of this

instrument. We evaluated the HBS-CHD to be a valid and responsive instrument for its use in research and clinical practice, although further research on the instrument's stability is required.

References

(1) Uzark K, VonBargen-Mazza P, Messiter E. Health education needs of adolescents with congenital heart disease. J Pediatr Health Care 1989;3(3):137-143.
(2) Staff J, Schulenberg JE, Maslowsky J, et al. Substance use changes and social role transitions: proximal developmental effects on ongoing trajectories from late adolescence through early adulthood. Dev Psychopathol 2010;22(4): 917-932.
(3) Chen P, Jacobson KC. Developmental trajectories of substance use from early adolescence to young adulthood: gender and racial/ethnic differences. J Adolesc Health 2012;50(2):154-163.
(4) Moons P, Budts W, Costermans E, et al. Pregnancy-related health behavior of women with congenital heart disease: room for behavioral change interventions. Congenit Heart Dis 2009; 4(5):348-355.
(5) Brickner ME, Hillis LD, Lange RA. Congenital heart disease in adults. First of two parts. N Engl J Med 2000;342(4): 256-263.
(6) Brickner ME, Hillis LD, Lange RA. Congenital heart disease in adults. Second of two parts. N Engl J Med 2000;342(5): 334-342.
(7) Sable C, Foster E, Uzark K, et al. Best Practices in Managing Transition to Adulthood for Adolescents With Congenital Heart Disease: The Transition Process and Medical and Psychosocial Issues: A Scientific Statement From the American Heart Association. Circulation 2011;123(13): 1454-1485.
(8) Gisle L, Hesse E, Drieskens S, et al. Operationele Directie Volksgezondheid en surveillance 2, Brussel Wetenschappelijk Instituut Volksgezondheid. Gezondheidsenquête, België 2008. Rapport II-Leefstijl en preventie. IPH/EPI REPORTS N° 2010/00. 2009.: https://www.wiv-isp.be/epidemio/ pinl/CROSPNL/HISNL/ his08nl /belangrijkste%20resultaten_NL.pdf
(9) Reid GJ, Webb GD, McCrindle BW, et al. Health behaviors among adolescents and young adults with congenital heart disease. Congenit Heart Dis 2008;3(1):16-25.
(10) Chen CW, Chen YC, Chen MY, Wang JK, Su WJ, Wang HL. Health-promoting behavior of adolescents with congenital heart disease. J Adolesc Health 2007;41(6): 602-609.
(11) Massin MM, Hovels-Gurich H, Seghaye MC. Atherosclerosis lifestyle risk factors in children with congenital heart disease. Eur J Cardiovasc Prev Rehabil 2007;14(2): 349-351.
(12) Van Deyk K, Moons P, Budts W. Health behaviour of adult patients with congenital heart disease. Eur J Cardiovasc Nurs 2005;4(1): 58.
(13) Babor TF, Higgings-Biddle JC, Saunders JB, et al. AUDIT The Alcohol Use Disorders Identification Test - Guidelines for Use in Primary Care. WHO/MSD/MSB/01.6a, 1-41. 2001. World Health Organization-Department of Mental Health and Substance Dependence. 2001, Geneva, Switzerland.
(14) Baecke JAH, Burema J, Frijters JER. A short questionnaire for the measurement of habitual physical activity in epidemiological studies. Am J Clin Nutr 1982;36: 936-942.
(15) Brener ND, Collins JL, Kann L, et al. Reliability of the youth risk behavior survey questionnaire. Am J Epidemiol 1995;141(6): 575-580.
(16) Center for Disease Control and Prevention-Division of Adolescent and School Health. Youth Risk Behavior Survey (YRBS) Standard 2011 High School Questionnaire Item Rationale.1-31. 2011: ttp://www.cdc.gov/healthyyouth/yrbs/pdf/questionnaire/ 2011_standard_itemrationale.pdf
(17) Florindo AA, Latorre MR, Santos EC, et al. Validity and reliability of the Baecke questionnaire for the evaluation of habitual physical activity among people living with HIV/AIDS. Cad Saude Publica 2006;22(3): 535-541.

(18) Kolbe LJ. An epidemiological surveillance system to monitor the prevalence of youth behaviors that most affect health. Health Education 1990;21: 44-48.
(19) Saunders JB, Aasland Og, Amundsen A, et al. Alcohol consumption and related problems among primary health care patients:WHO collaborative project on early detection of persons with harmful alcohol consumption-I. Addiction 1993;88(3): 349-362.
(20) Saunders JB, Aasland OG, Babor TF, et al. Development of the Alcohol Use Disorders Identification Test (AUDIT): WHO Collaborative Project on Early Detection of Persons with Harmful Alcohol Consumption-II. Addiction 1993;88(6): 791-804.
(21) Vanheusden K. Mental health problems and barriers to service use in Dutch young adults. Rotterdam, The Netherlands; Erasmus University, 2008.
(22) Reid GJ, Siu SC, McCrindle BW, et al. Sexual behavior and reproductive concerns among adolescents and young adults with congenital heart disease. Int J Cardiol 2008; 125(3):332-338.
(23) Godin G, Shephard RJ. A simple method to assess exercise behavior in the community. Can J Appl Sport Sci 1985;10: 141-146.
(24) Durnin JVGA, Passmore R. Energy, work and leisure. 1ed. London: Heinemann Educational Books Ltd.; 1967.
(25) American Educational Research Association, American Psychological Association, National Council on Measurement in Education, Joint Committee on Standards for Educational and Psychological Testing. Standards for Educational and Psychological Testing. Washington DC: American Educational Research Association, 1999.
(26) Davis LL. Instrument review: Getting the most from a panel of experts. Appl Nurs Res 1992;5: 194-197.
(27) Grant JS, Davis LT. Selection and use of content experts in instrument development. Res Nurs Health 1997;20: 269-274.
(28) Polit DF, Beck CT, Owen SV. Is the CVI an acceptable indicator of content validity? Appraisal and recommendations. Res Nurs Health 2007;30(4): 459-467.
(29) Waltz CF, Strickland O, Lenz ER, et al. Measurement in Nursing and Health Research. 3rd ed. New York: Springer Publishing Co., 2005.
(30) Cicchetti DV, Sparrow S. Developing criteria for establishing interrater reliability of specific items: Application to assessment of adaptive behavior. Am J Ment Defic 1981; 86: 127-137.
(31) Fleiss JL. Statistical Methods for Rates and Proportions 2 ed. New York: Chichester: Brisbane: John Wiley & Son; 1981.
(32) Health-risk behaviors among persons aged 12-21 years- United States, 1992. From the Centers for Disease Control and Prevention. Health-risk behaviors among persons aged 12-21 years-United States, 1992. JAMA 1994;271(17): 311.
(33) Philippaerts RM, Westerterp KR, Lefevre J. Double labelled water validation of three physical activity questionnaires. J Sport Med 1999;20: 284-289.
(34) Philippaerts RM, Westerterp KR, Lefevre J. Comparison of two questionnaires with a tri-axial accelerometer to assess physical activity patterns. J Sport Med 2001;22: 34-39.
(35) Goodwin LD, Leech NL. Changing conceptions of measurement validity: an update on the new standards. Measurement and Evaluation in Counseling and Development 2003; 36: 181-191.
(36) Knapp TR, Kimble LP, Dunbar SB. Distinguishing between the stability of a construct and the stability of an instrument in trait/state measurement. Nurs Res 1998;47(1): 60-62.
(37) Heise DR. Separating Reliability and Stability in Test-retest Correlation. Am Sociol Rev 1969;34(1):93-101.
(38) Liang MH. Longitudinal construct validity: Establishment of clinical meaning in patient evaluative instruments. Med Care 2000;38(9):84-90.
(39) Husted JA, Cook RJ, Farewell VT, eta l. Methods for assessing responsiveness: A critical review and recommendations. J Clin Epidemiol 2000;53(5):459-468.
(40) Guyatt G, Walter S, Norman G. Measuring change over time: Assessing the usefulness of evaluative instruments. J Chronic Dis 1987;40(2):171-178.

(41) Mitchell SC, Korones SB, Berendes HW. Congenital heart disease in56,109 births. Incidence and natural history. Circulation 1971;43(3): 323-332.
(42) Montoye HJ, Kemper HCG, Saris WHM, et al. Measuring physical activity and energy expenditure. In: Champaign IL, editor. Human kinetics. 1996: p. 191.
(43) Pereira MA, Fitzergerald SJ, Gregg EW, et al. A collection of physical activity questionnaires for health-related research. Med Sci Sports Exerc 1997;29 (6(Suppl.1)):S1-S205.
(44) Dillman D. Mail and other self-administered questionnaires. Handbook of survey research. New York: Academic Press; 1983.
(45) Luyckx K, Goossens E, Van Damme C, et al. Identity formation in adolescents with congenital cardiac disease: a forgotten issue in the transition to adulthood. Cardiol Young 2011;21(4): 411-420.
(46) Rickham PP. Human experimentation. Code of ethics of the world medical association. Declaration of Helsinki. Br Med J 1964; 2(5402): 177.
(47) European Monitoring Centre for Drugs and Drug Addiction. Thematic papers: Hallucinogenic mushrooms: an emerging trend case study. ISBN 92-9168-249-7, 1-33. 2006.: http:// www.emcdda.europa.eu/publications/thematic-papers/mushrooms
(48) World Health Organization. Global recommendations on physical activity for health. ISBN 978 924 159 997, 1-58. 2010, Geneva, Switzerland
(49) Habib G, Hoen B, Tornos P, et al. Guidelines on the prevention, diagnosis, and treatment of infective endocarditis (new version 2009): the Task Force on the Prevention, Diagnosis, and Treatment of Infective Endocarditis of the European Society of Cardiology (ESC). Endorsed by the European Society of Clinical Microbiology and Infectious Diseases (ESCMID) and the International Society of Chemotherapy (ISC) for Infection and Cancer. Eur Heart J 2007;30(19): 2369-2413.
(50) Wilson W, Taubert KA, Gewitz M, et al. Prevention of infective endocarditis: guidelines from the American Heart Association: a guideline from the American Heart Association Rheumatic Fever, Endocarditis, and Kawasaki Disease Committee, Council on Cardiovascular Disease in the Young, and the Council on Clinical Cardiology, Council on Cardiovascular Surgery and Anesthesia, and the Quality of Care and Outcomes Research Interdisciplinary Working Group. Circulation 2007;116(15): 1736-1754.
(51) Busuttil NA, Mooney G, El Bahannasawy E, et al. The dental health and preventative habits of cardiac patients attending the Royal Hospital for Sick Children Glasgow. Eur Arch Paediatr Dent 2006;7(1):23-30.
(52) Duval X LC. Prophylaxis of infective endocarditis: current tendencies, continuing controversies. Lancet Infect Dis 2008;8: 225-232.
(53) Forner L, Larsen T, Kilian M, et al. Incidence of bacteremia after chewing, tooth brushing and scaling in individuals with periodontal inflammation. J Clin Periodontol 2006;33: 401-407.
(54) Lockhart PB, Loven B, Brennan MT, et al. The evidence base for the efficacy of antibiotic prophylaxis in dental practice. J Am Dent Assoc 2007;138(4):458-474.
(55) Lockhart PB, Brennan MT, Thornhill M, et al. Poor oral hygiene as a risk factor for infective endocarditis-related bacteremia. J Am Dent Assoc 2009;140(10):1238-1244.
(56) Roberts GJ. Dentists are innocent! 'Everyday' bacteremia is the real culprit: a review and assessment of the evidence that dental surgical procedures are a principal cause of bacterial endocarditis in children. Pediatr Cardiol 1999;20: 317-325.
(57) Crasta K, Daly CG, Mitchell D, et al. Bacteraemia due to dental flossing. J Clin Periodontol 2009;36: 323-332.
(58) Storm BL, Abrutyn E, Berlin JA, et al. Risk factors for infective endocarditis: oral hygiene and nondental exposures. Circulation 2000;102: 2842-2848.
(59) Norusis MJ. Measuring scales: Reliability analysis examples. SPSS Professional Statistics 7.5. Chicago: SPSS Inc., 1997, pp.103-111.
(60) Warnes CA, Liberthson R, Danielson GK, et al. Task force 1: The changing profile of congenital heart disease in adult life. J Am Coll Cardiol 2001;37(5):1170-1175.

(61) Saunders JB, Aasland OG, Amundsen A, et al. Alcohol consumption and related problems among primary health care patients: WHO collaborative project on early detection of persons with harmful alcohol consumption-I. Addiction 1993;88(3):349-362.

Appendix- Figure 7.3: Health Behavior Scale –Congenital Heart Disease - English (USA).

Health behaviour

This questionnaire is about your health behavior. ***Colour*** the correct answer black.

Only ***1 answer per question*** please.

1. **Do you consume alcohol from time to time? (by alcohol is meant: beer, wine, liquor, coolers,...)**

☐ No (Proceed to question 4)

☐ Yes

If yes, how often?

- once a month or less
- 2 to 4 times a month
- 2 to 3 times a week
- 4 or more times a week

2. **When consuming alcohol, how many glasses do you have on average?**
   - 1 to 2
   - 3 to 4
   - 5 to 6
   - 7 to 9
   - 10 or more

3. **How often do you drink 6 glasses or more on one occasion?**
   - Never
   - Less than once a month
   - Monthly
   - Weekly
   - Daily or almost every day

4. Do you smoke cigarettes occasionally or regularly?

☐ No

(Proceed to question 7)

☐ Yes

5. During the last 30 days, on how many days did you smoke cigarettes?

- ○ 1 to 2 days
- ○ 3 to 5 days
- ○ 6 to 9 days
- ○ 10 to 19 days
- ○ 20 to 29 days
- ○ on all 30 days

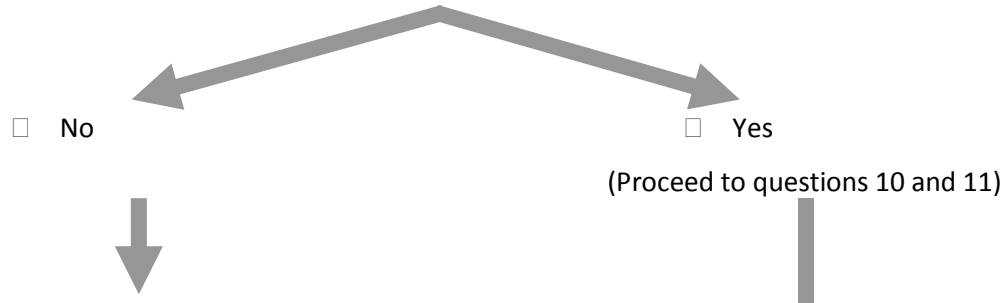
6. During the last 30 days, on the days you smoked, how many cigarettes did you smoke a day?

- ○ 1 cigarette or less a day
- ○ 2 to 5 cigarettes a day
- ○ 6 to 10 cigarettes a day
- ○ 11 to 20 cigarettes a day
- ○ more than 20 cigarettes a day

7. How often, in the last 12 months, did you take the following drugs?

		Never	once a month or less	2 to 4 times a month	2 times or more a week
a.	Cannabis (marihuana, hash)	○	○	○	○
b.	XTC (ecstasy)	○	○	○	○
c.	Cocaine	○	○	○	○
d.	Hallucinogenic mushrooms	○	○	○	○
e.	Speed	○	○	○	○
f.	Sleeping pills, sedatives or tranquillizers	○	○	○	○
g.	Other (illicit)drugs:	○	○	○	○

8. Have you been to the dentist in the past year?



☐ No

☐ Yes

(Proceed to questions 10 and 11)

9. If not, when did you last go to the dentist?

- I never go to the dentist
- 1-2 years ago
- 2-3 years ago
- more than 3 years ago

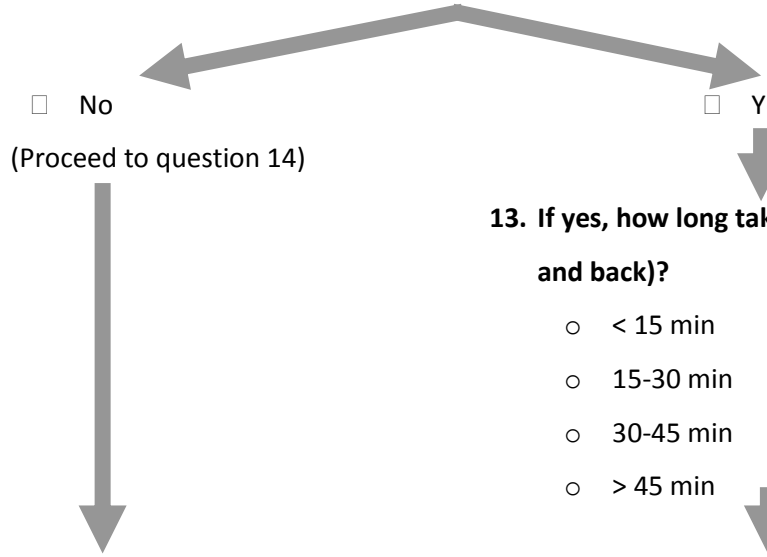
10. How often do you brush your teeth?

- I don't brush my teeth
- I brush my teeth every now and then
- once a day
- twice a day
- 3 times a day
- more than 3 times a day

11. **How often do you floss your teeth?**

- I don't floss my teeth
- I floss my teeth every now and then
- once a day
- twice a day
- 3 times a day
- more than 3 times a day

12. Do you regularly walk or cycle to school or to your place of work?



☐ No

(Proceed to question 14)

☐ Yes

13. If yes, how long take it take by bike or on foot (there and back)?

- o < 15 min
- o 15-30 min
- o 30-45 min
- o > 45 min

14. Do you regularly practice a sport? (this includes school sports but <u>NOT</u> the bike ride or walk to school or your work place

☐ No (the questionnaire stops here; go to the next page)

☐ Yes

15. During a 7-day week, how many hours of the following physical activities do you do?

a. <u>Sport at school</u>, during P.E. lessons or other sports periods

☐ hours/week

b. Sports or activities that are <u>very physically demanding, which increase your pulse</u> (e.g. football, a long run, basketball, handball, korfball, squash, rowing, rugby, hockey, spinning, Thai boxing, kickboxing, cycle racing, rope-skipping, mountain biking, tennis,...)

☐ hours/week

c. Sports or activities that are <u>moderately physically demanding</u> and where, afterwards, you <u>don't feel exhausted or worn out</u> (e.g. jogging, volleyball, swimming up and down, ballet, dancing, judo, karate, athletics, badminton, baseball, fitness classes, horse riding, wall climbing,...)

☐ hours/week

d. d. Sports or activities with minimal physical effort or gentle exertions (e.g. billiards, bowling, darts, golf, playing cards, yoga, fishing,...)

☐ hours/week

8 IMPACT OF STRUCTURED EDUCATION ON DISEASE-RELATED KNOWLEDGE LEVEL IN YOUNG PEOPLE WITH CONGENITAL HEART DISEASE

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ABSTRACT

Background: Patients with congenital heart disease (CHD) have poor understanding of their heart condition, treatment and prevention of complications. To improve their level of health-related knowledge, a structured education program was implemented at the adult congenital heart disease program. This study aimed (a) to evaluate the level of knowledge of patients who received structured CHD education as compared to patients who did not receive this education; (b) to explore if the provision of structured education is an independent determinant of knowledge; and (c) to evaluate if patients who received structured education reached the educational target (>80% correct answers).

Methods and results: A total of 317 patients were included: 226 in the education group, and 91 in the comparison group. Knowledge was assessed using the 'Leuven Knowledge Questionnaire for Congenital Heart Disease'. The mean total knowledge score in the education group (57%) was significantly higher as compared to the comparison group (43%) (p<0.001). However, only 24 patients (11%) in the education group reached the educational target of the program. After adjusting for patient's age, educational level and disease complexity, hierarchical multivariable linear regression analysis showed that the provision of structured CHD education was an independent determinant of higher levels of knowledge.

Conclusion: A structured education program was associated with a higher level of knowledge. However, the educational target for sufficient knowledge was reached in a very limited number of patients. Hence, continuous efforts in educating patients and developing alternative education methods are needed.

Introduction

A significant proportion of patients diagnosed with congenital heart disease (CHD) remain at risk for developing cardiac complications such as atrial arrhythmias, infective endocarditis and congestive heart failure (1,2). A good understanding of the heart condition, treatment and components of heart-healthy behavior is necessary to prevent these potential complications (3,4). Limited knowledge of patients might result in health-compromising behaviors or result in unseemly restrictions influencing the well-being of patients. A good understanding of the impact of the heart condition on daily living is fundamental especially for adolescents as they are prepared to become responsible for their healthcare and self-management in adult life (5).

Studies that comprehensively assessed the level of knowledge of adults (3,6) and adolescents with CHD (5) are scarce. Previous studies predominantly evaluated selected aspects of knowledge in these patients such as the name and location of the heart defect (7-13), prevention of endocarditis (8,10-12,14), inheritance (15), risk of pregnancy (11), restrictions to physical activity (16), use of contraceptives (17) and medication for anti-coagulation (18). These studies demonstrated that adolescents and adults with CHD have poor to moderate understanding of their heart condition (3,5-13), aspects of self-care activities preventing endocarditis (3,5,6,8-12,14,18), inheritance of CHD and reproductive issues(3,6,15,17), and safe levels of physical activity (3,5,9,16,18). The comparability of these cross-sectional studies is however limited since the level of knowledge was assessed using different methods: by interview (7,14); drawing techniques (13); or structured questionnaires (3,5,6,8-10,12,15,16,18-21). However, several studies that used questionnaires, had limited or no data that supported their validity or reliability of the questionnaires (8-10,12,15,16,18).

To enhance the level of knowledge in patients, repetitive and structured educational interventions are recommended (11,12,19,22). Since provision of patient education is a time-consuming activity, requiring the investment of sufficient time, space and trained personnel, studies evaluating the effectiveness of patient education are highly needed (12,23). However, the evidence supporting the effectiveness of structured education programs is currently lacking. A preliminary pre-post study showed a promising 10% increase of knowledge levels in 31 adults with CHD after the implementation of a structured education program (19). Since no study comparatively investigated the effectiveness of educational efforts, this study aimed (a) to compare the level of knowledge between patients with CHD who received structured education and a comparison group of patients who did not receive prior structured education; (b) to explore if the provision of structured education

was an independent determinant of higher levels of knowledge; and (c) to evaluate if patients who received education reached the educational target of the education program.

Methods

Study population

In this descriptive, cross-sectional study, patients with CHD were recruited at the outpatient clinic for adults with CHD. Patients were eligible if they had a structural heart defect, were literate, Dutch speaking, and gave verbal informed consent. Patients were excluded if they had learning disabilities, if they were not in regular follow-up at the pediatric cardiology or ACHD program of the hospital. Patients who were newly diagnosed as adults with an atrial septal defect or foramen ovale after a cryptogenic stroke were excluded since the congenital cardiac condition was diagnosed secondary to the stroke.

Participants were recruited on preselected dates during a 13-month period from January 2006 to February 2007. Overall, 493 patients were planned for an outpatient visit on these selected dates, 353 (72%) of which met the inclusion criteria and were therefore asked to partake in the study. Six patients refused to participate due to a lack of interest or time constraints. Thirty patients were excluded because of practical reasons, e.g. did not bring their glasses; physician was waiting; etc. Thus, 317 (90%) of the eligible patients participated. Participants were divided into two groups: (a) patients who previously had an outpatient visit at the ACHD clinic and therefore received at least once structured patient education, and (b) patients who transferred directly from pediatric cardiology or other adult services and had their very first ACHD visit. These latter patients comprised the comparison group since they did not receive structured education before. The education group consisted of 226 patients with a median age of 27.5 years; 54% were male. The comparison group consisted of 91 patients with a median age of 17 years; 53% were male. Demographic and clinical characteristics of both groups are detailed in **Table 8.1**.

Table 8.1: Demographic and clinical characteristics of 317 patients with congenital heart disease

Variable	Education group (n=226)	Comparison group (n=91)
Sex		
Male	123 (54%)	48 (53%)
Female	103 (46%)	43 (47%)
Median age (in years)	27.5 (Q_1=23; Q_3=34) (Range: 16-60)	17 (Q_1= 16; Q_3=18) (Range: 15-32)
Complexity of CHD		
Simple	40 (18%)	32 (35%)
Moderate	152 (67%)	48 (53%)
Complex	34 (15%)	11 (12%)
Marital status		
Unmarried (living with parents)	98 (43%)	87 (96%)
Married or living together	18 (8.0%)	3 (3%)
Living alone	110 (49%)	1 (1%)
Highest educational level		
Vocational high school	60 (28%)	29 (31%)
Technical high school	35 (16%)	32 (34%)
High school/College/University	123 (56%)	30 (32%)
Primary medical diagnosis		
Tetralogy of Fallot	47 (21%)	4 (4%)
Aortic valve disease	44 (19%)	11 (12%)
Coarctation of the aorta	29 (13%)	12 (13%)
Ventricular septal defect	24 (11%)	26 (29%)
Transposition of the great arteries	14 (6%)	5 (5%)
Pulmonary valve disease	11 (5%)	11 (12%)
Other	57 (25%)	22 (24%)

Responsible for the daily management of care		
Parents	-	1 (1%)
Patient	210 (93%)	23 (25%)
Patient and parents	16 (7%)	67 (74%)
History of infective endocarditis	9 (4%)	0 (0%)
History of pregnancies (for women only)	36 (35%)	0 (0%)
Contraception (for women only)		
Birth control pill	66 (66%)	19 (43%)
Other methods	10 (10%)	1 (2%)
No contraception	24 (24%)	24 (55%)

CHD = Congenital Heart Disease

Setting and Intervention

The division of Congenital Cardiology, established in 1960, is part of the University Hospitals of Leuven in Belgium and contains two programs: pediatric cardiology and the ACHD program. To date, more than 35,000 individuals have been examined and/or treated in this center. In 1995, a dedicated ACHD program with a different provider team was established. At this hospital, young people with CHD are transferred from pediatric cardiology to the ACHD program when they reach the age of 16, unless they are medically unstable. This transfer is not preceded by structured provision of information or instruction as part of a formal transition program.

According to the complexity of the heart defect, patients are planned to have an outpatient visit on a regular basis (24-26). For patients diagnosed with a simple heart lesion, outpatient visits to check the patient's cardiac condition, are planned every 3-5 years either in a non-specialized setting or at a shared care facility (24,25). In patients with a moderately severe condition, check-ups are recommended every 1-2 years preferably done at specialist centers (24). Patients who have a complex heart lesion are advised to have a follow-up visit at an ACHD program every 6-12 months (24).

A typical outpatient visit of an adult patient to the ACHD program starts with the registration of an electrocardiogram (ECG) by a nurse. Then patients will meet one of the nurses of the ACHD advanced practice nursing (APN) team to discuss their subjective condition, experienced symptoms, lifestyle issues and questions they have. Furthermore, structured patient education is provided verbally during this session. This education program was based on the framework of educational assessment in CHD (27) and provides information about the heart defect, the current treatment, the need for regular follow-up, characteristics and prevention of endocarditis, physical and vocational lifestyle issues, inheritability of the heart defect, potential risk of pregnancy, contraception, sexuality and traditional cardiovascular lifestyle issues. Education is provided to patients on an individual basis during an outpatient visit and takes 15-30 min. Topics are discussed repeatedly during subsequent outpatient visits to facilitate retention of the provided information. During the education session, a computerized intake form/checklist is used by the nursing team to guarantee that information is provided in a structured and systematic way. Finally, the last part of the outpatient visit entails a visit to the congenital cardiologist who performs the medical check-up using clinical examination, echocardiography, or other diagnostic tools.

To date, two advanced practice nurses and three specialized nurses are on the staff in the ACHD program, corresponding with 1.73 full-time equivalents. With the current staffing, education

can systematically be provided during each outpatient visit, which resulted in 2,528 patients who received education in the year 2012.

Variables and measurement

Demographic data were gathered during a patient interview. Clinical variables (primary diagnosis, disease complexity as defined by Task Force 1 of the 32nd Bethesda conference (28), history of endocarditis, history of pregnancy, prescribed contraception) were collected from patients' medical records. Assessment of the level of knowledge of all participants, irrespective of the group to which they were assigned, was performed using the Leuven Knowledge Questionnaire for Congenital Heart Disease (LKQCHD) (3,29). This instrument was developed and refined by Moons and coworkers as a comprehensive questionnaire assessing knowledge in patients with CHD. A revised version of the LKQCHD was developed in 2009 comprising 34 questions for female and 31 questions for male patients (29). The LKQCHD covers five domains: (a) knowledge of the heart defect and treatment, (b) knowledge of the prevention of complications, (c) knowledge of physical activities, (d) knowledge of sexuality and heredity, and (e) knowledge of contraception and pregnancy planning, and has been described in detail elsewhere (3). Expert review, pilot testing, and confirmation of theoretically derived hypotheses provided evidence for the content and construct validity of the LKQCHD (3,29).

Procedure

A nurse from the ACHD nursing team explained the purpose and procedure of the study to all patients prior to their scheduled outpatient visit at the ACHD clinic. After receiving verbal consent, the nurse gave the patients instructions on how to fill out the LKQCHD while they were sitting in the waiting room. To ensure that the patients filled out the questionnaire independently, the nurse asked relatives not to help. External sources (e.g., internet) could not be consulted. After completion of the LKQCHD, the nurse checked whether all items were filled out and asked for clarification if necessary. The researcher evaluated the patients' answers as 'correct,' 'does not know,' 'incorrect,' or 'incomplete.' The study was approved by the Institutional Review Board of the University Hospitals Leuven, Belgium.

Statistical analysis

Statistical analyses were performed using SPSS, version 17.0 (SPSS Inc., Chicago, Illinois, USA). Data derived from the LKQCHD were analyzed at two levels: item level and patient level. At the item level, knowledge variables were dichotomized as 'correct' or 'incorrect' (i.e. includes incomplete, does not know, and incorrect responses), and the proportion of patients that gave a

correct answer was calculated. At the patient level, a total knowledge score was calculated by computing the percentage of correct answers per patient. The educational target for individual patients in this education program was set at an overall knowledge score of ≥80% based on a cut-off used in previous studies (3,5). Descriptive statistics for nominal variables were expressed in absolute numbers and percentages. Continuous data were presented as means and standard deviations if data were normally distributed. To test differences in overall knowledge score between the education and comparison group, an unpaired *t*-test was used. The effect size was determined by calculating the Cohen's *d*. Effect sizes were categorized as a small (0.2–0.5); moderate (0.5–0.8); or large difference (>0.8) (30).

To explore if the level of knowledge was independently determined by the structured education provided, a hierarchical multivariable linear regression analysis (enter method) was performed, adjusting for confounding factors, potential differences between the groups, and interaction effects. Based on empirical evidence, three potential confounders were included in this analysis: age (3,7,9,10,12), level of education (3,9,12,15), and complexity of disease (3). The assumptions of normality and linearity of residuals, and absence of multicollinearity were met. Furthermore, to test the robustness of this model a matched (1:1) control analysis was performed, in which the difference in knowledge levels was directly tested between pairs of patients, matched on the confounders emerging from the multivariable model. All tests were two-sided, and the level of significance was set at $p < 0.05$.

Results

Comparison of the level of knowledge

Patients who previously received education displayed adequate knowledge in 8 of 34 (24%) questions (>80% respondents answered correctly); moderate knowledge in 11 of 34 (32%) questions (50-80% respondents answered correctly); and poor knowledge in 15 of 34 (44%) questions (<50% respondents answered correctly).

Patients who did not receive prior education (comparison group) displayed adequate knowledge in 5 of 34 (15%) questions; moderate knowledge in 7 of 34 (20%) questions; and poor knowledge in 22 of 34 (65%) questions. The proportion of patients providing a correct answers for all items of the LKQCHD was consistently higher in the education group, with the exception of items 5 and 17 (**Table 8.2**).

Patients in the education group had a significantly higher mean total knowledge score (57%), as compared to the comparison group (43%)(t=8.737; p<0.001). The difference between the scores of the two groups was not only statistically different, but also clinically meaningful as shown by the calculation of the Cohen's d (d = 1.24).

Exploration of independent determinants of knowledge

In order to correct for confounding factors and for differences between the groups, a hierarchical multivariable linear regression analysis was conducted. This analysis revealed that, after adjusting for age, educational level, and disease complexity (block 1), structured patient education (block 2) was significantly associated with the total knowledge score (**Table 8.3**). This model demonstrated that a higher educational background and a higher disease complexity were significant correlates of a higher level of knowledge. This means that structured patient education was a significant factor in explaining improved levels of patient knowledge, over and above the impact of educational background and disease complexity. The knowledge score was not related to patients' age, suggesting that knowledge in this population is not time-dependent. Moreover, no interaction effect between age and structured education was observed. The final regression model explained 28% of the variance for the level of knowledge (**Table 8.3**).

The matched control analysis, comparing the level of knowledge of 85 pairs of patients matched on educational background and disease complexity showed that the knowledge score of the education group was significantly higher as compared to the matched controls (t=-4.474; p<0.001). This displays the robustness of the model and provides additional support for the effectiveness of structured patient education.

Proportion of patients reaching the educational target

Although the level of knowledge was significantly higher in the education group, the educational target of the program, defined as a total knowledge score of ≥80, was only achieved in 24 (11%) patients of the education group.

Table 8.2: Proportion of correct answers in patients who previously received education (education group) and those without prior education (comparison group)

Question	Education group (n=226)	Comparison group (n=91)
1. What is the name of your heart defect?	150/226 (66%)	41/91 (45%)
2. Describe or indicate on the diagram where your heart defect is located.	109/226 (48%)	25/91 (27%)
3. How often do you have to come to the clinic for follow-up for your conenital heart disease?	206/226 (93%)	71/91 (78%)
4. What is the main purpose of the follow-up?	128/226 (57%)	42/91 (46%)
5. How has your heart condition been treated to date?	177/226 (78%)	77/91 (85%)
6. If you are on drug treatment, give the name, dose, schedule, reason or function, most important side effects, and interactions with other drugs or foods.	59/89 (66%)	8/15 (53%)
7. If you experience side effects from your drugs, does this mean you should stop taking them?	137/226 (61%)	37/91 (41%)
8. Do you have to follow a diet? If you answer yes, please indicate the type of diet.	218/226 (97%)	86/91 (94%)
9. Mark all symptoms that may occur if your heart condition deteriorates and for which you have to contact your cardiologist.	71/226 (31%)	8/91 (9%)
10. If the (congenital) cardiologist informs you that everything is alright, does that mean that you do not need further follow-up?	209/226 (93%)	79/91 (87%)
11. What is endocarditis?	96/226 (43%)	19/91 (21%)
12. What is the most typical sign or symptom of endocarditis?	44/226 (20%)	1/91 (1%)
13. Can you only get endocarditis once in your lifetime?	88/226 (39%)	13/91 (14%)

Question	Education group (n=226)	Comparison group (n=91)
14. Do the following risk factors contribute to the onset of endocarditis?		
Needle contamination (drug users)	95/226 (42%)	18/91 (20%)
Smoking	48/226 (21%)	7/91 (8%)
Bacteria from skin infections	90/226 (40%)	10/91 (11%)
Dental abscesses	133/226 (59%)	20/91 (22%)
Sexual activity	91/226 (41%)	22/91 (24%)
Poor nail and skin care	72/226 (32%)	7/91 (8%)
Body piercing and tattooing	85/226 (38%)	14/91 (15%)
15. As you have a congenital heart disease, should you take antibiotics immediately (without consulting a physician) if you gave a temperature?	191/226 (85%)	71/91 (78%)
16. Should you have a dental check-up at least once a year?	209/226 (93%)	76/91 (84%)
17. Should you take antibiotics before every visit to the dentist?	152/226 (67%)	71/91 (78%)
18. Do bleeding gums need extra attention?	200/226 (89%)	68/91 (75%)
19. Should you clean your teeth at least once a day?	220/226 (97%)	87/91 (96%)
20. Is smoking more harmful for patients with congenital heart disease than for other people?	32/226 (14%)	7/91 (8%)
21. Is consuming alcohol 3 times a day more harmful for patients with congenital heart disease than for other people?	60/226 (27%)	22/91 (24%)
22. Can you take part in competitive sports (regional or national) requiring daily training?	125/226 (55%)	35/91 (39%)
23. Should you choose an occupation that is not too physically demanding, as you should be careful not to overexert yourself?	173/226 (77%)	68/91 (75%)
24. In terms of sexual physical effort, can you do all what you feel you are capable of doing?	205/226 (91%)	71/91 (78%)
25. What is the chance that your children will have congenital heart disease?	66/226 (29%)	18/91 (20%)
26. Which contraceptives are the most advisable for you to use in light of your congenital heart disease? (only for women)		
contraceptive pill	65/103 (63%)	15/43 (35%)
Intrauterine device	29/103 (28%)	6/43 (14%)
27. Are you at risk for deterioration or complications during pregnancy? (only for women)	51/103 (50%)	5/43 (12%)

Questions in this table replicate the Leuven Knowledge Questionnaire for Congenital Heart Disease (LKQCHD)

Table 8.3: Hierarchical multivariable linear regression analysis on the effect of education on total knowledge score in 317 patients with congenital heart disease, adjusted for age, education background, and disease complexity.

Variable	Estimate	Standard error	Standardized estimate	p-value
Intercept	9.28	4.27	-	0.031
Education (vs. no prior education)	11.45	2.20	0.31	<0.001
Age	0.03	0.11	0.02	0.790
Educational level	6.64	0.94	0.35	<0.001
Disease complexity	4.65	1.36	0.17	0.001

Adjusted R^2= 0.277

Discussion

This study showed that patients who partook in a structured education program were significantly more knowledgeable about their heart defect, its treatment, and various preventive measures than patients who did not receive structured education. Furthermore, this difference in knowledge levels of both groups was found to be large and clinically relevant.

Hierarchical multivariable regression analysis showed that the provision of structured patient education was a significant and independent predictor of higher levels of knowledge in patients. Additionally, higher levels of educational background and a more complex heart defect were identified as significant predictors of increased levels of knowledge in this study. Higher levels of knowledge might be expected in patients with more complex CHD, since these patients are recommended to have an outpatient visit at least once a year. Therefore, these patients will receive structured education more frequently and as a result information might be better retained. Furthermore, this study confirmed previous study results that indicated a positive correlation between knowledge of CHD and higher educational levels(3,9,12,15,29). This finding stresses the importance of providing more attention to patients with lower educational levels because they might need more time or repetition in order to understand the information and lifestyle instructions. As in line with previous studies in adolescents (3,10,12) and adults (7) with CHD, the level of knowledge was found to be independent of age. Since there were significant differences in the size of the two groups and in the characteristics of patients belonging to either the education or comparison group, a matched case control analysis was performed. This analysis created 85 pairs of patients matched on their educational background and CHD complexity. Since age was not identified as a significant

determinant of knowledge, patients were not matched with a control on their respective age. In sum, this study was the first to show that structured patient education by an advanced practice ACHD nursing team is effective in improving patient's knowledge about their heart condition, treatment, lifestyle issues and health-promoting behaviors.

Nonetheless, this study revealed that patients with CHD who received structured patient education still have a relatively low level of knowledge on multiple aspects of their condition since only 1 in 10 patients achieved the predefined educational target. Analysis of the responses of patients in the education group showed that most patients still had a poor understanding of the definition, symptoms, and risk factors of endocarditis; the signs of deterioration of the heart condition; inheritance of the defect; the risk of pregnancy; the anatomy of the lesion; and the suitability of intrauterine devices as contraceptives. Hence, the educational efforts currently implemented in the ACHD program of the hospital were not capable of achieving the anticipated educational target. This result might be disappointing, given the investment of supplementary resources which are needed to systematically and repetitively provide education to these patients. However, previous studies showed that retention of health-related information is generally low in patients (31-33). Therefore, complementary or alternative forms of patient education should be explored. To date, education was provided verbally in this ACHD program. Alternative forms of education could be provided by means of written information (booklets), video education, or interactive computer programs such as digital game-based learning (34-36). This latter type of education seems to be a promising method to educate young people with a chronic condition. Rönning and colleagues performed an initial evaluation of the implementation of an Education and Psychosocial Support (EPS) model (21). This model comprised three components: medical consultation by an ACHD cardiologist, computer-based education, and psychosocial support provided by a multidisciplinary team. A 3-month follow-up of 55 adults who received this type of education showed an increase in self-perceived levels of knowledge. Unfortunately, this pre-post study did not objectively assess the impact of this model on the level of knowledge through the use of a questionnaire (21).

Methodological issues

One must consider some methodological limitations inherent to this study when interpreting these findings. First, since this cross-sectional study was conducted in a single outpatient clinic of a university hospital in Belgium, study results have limited generalizability. However, this single-center approach was critical to yield a relatively large sample size, low attrition rate, and no missing data. An RCT-design is generally used to test the effectiveness of an intervention in patients. Since structured education is a standard element of the current care provision, it would be unethical to withhold part of the patients of this intervention. Therefore, this study used a non-randomized comparison design using a convenience comparison group reflecting the day-to-day practice. Furthermore, knowledge about CHD was measured using the LKQCHD, an instrument for which previous studies provided evidence to support the content validity and validity based on relations with other variables (3,29). However, additional aspects of reliability and validity of this instrument remain subject to scrutiny (29). Third, the LKQCHD questionnaire gives the same weighting to all items although it might be argued that some items, e.g. prevention of endocarditis, would be of more importance to a subset of patients than to others. For the purpose of this study, a total knowledge score was calculated by simply computing the percentage of correct answers. Future studies should evaluate whether calculation of a summary score based on weighing factors assigned to items with a higher relevance would be meaningful and relevant. Fourth, although all nurses of the APN team were trained to provide structured education in a standardized way, variation in the education provided by these nurses could not be excluded. This variation might result in bias, but is a reflection of the real-life clinical practice.

Conclusion

This study was the first to support the effectiveness of structured patient education provided by an advanced practice nursing team in improving the level of knowledge of adolescents and adults with CHD using a comparative study design. Unfortunately, the educational target of this program was only reached by a limited number of educated patients. Therefore, clinicians and researchers should continue their efforts in teaching patients and in developing alternative education methods.

References

(1) Perloff JK. Residua and sequelae: a perspective. In: Perloff JK, Child JS, editors. Congenital heart disease in adults. Philadelphia: WB Saunders, 1998, pp. 303-315.
(2) Somerville J. Management of adults with congenital heart disease: an increasing problem. Annu Rev Med 1997; 48:283-293.
(3) Moons P, De Volder E, Budts W, et al. What do adult patients with congenital heart disease know about their heart disease, treatment, and prevention of complications? Heart 2001; 86:74-80.
(4) Gochman D. Handbook of health behavior research. New York: Plenum Press, 1997.
(5) Van Deyk K, Pelgrims E, Troost E, et al. Adolescents' understanding of their congenital heart disease on transfer to adult-focused care. Am J Cardiol 2010; 106:1803-1807.
(6) Chessa M, De Rosa G, Pardeo M, et al. Illness understanding in adults with congenital heart disease. Ital Heart J 2005; 6:895-899.
(7) Veldtman GR, Matley SL, Kendall L, et al. Illness understanding in children and adolescents with heart disease. Heart 2000; 84:395-397.
(8) Cetta F, Podlecki DC, Bell TJ. Adolescent knowledge of bacterial endocarditis prophylaxis. J Adolesc Health 1993; 14:540-542.
(9) Ferencz C, Wiegmann FL Jr, Dunning RE. Medical knowledge of young persons with heart disease. J Sch Health 1980; 50:133-136.
(10) Dore A, de Guise P, Mercier L-A. Transition of care to adult congenital heart centres: What do patients know about their heart condition? Can J Cardiol 2002; 18:141-146.
(11) Cetta F, Warnes CA. Adults with congenital heart disease: patient knowledge of endocarditis prophylaxis. Mayo Clin Proc 1995; 70:50-54.
(12) Kantoch MJ, Collins-Nakai RL, Medwid S,et al. Adult patients' knowledge about their congenital heart disease. Can J Cardiol 1997; 13:641-645.
(13) Wang Q, Hay M, Clarke D, et al. Adolescents' drawings of their cardiac abnormality. Cardiol Young 2011; 21:556-561.
(14) Vogel M, Knirsch W, Lange PE. Severe complications caused by inattention to endocarditis prevention during dental procedures in adults with congenital heart abnormalities. Dtsch Med Wochenschr 2000; 125:344-347.
(15) van Engelen K, Baars MJH, van Rongen LT, et al. Adults with congenital heart disease: Patients' knowledge and concerns about inheritance. Am J Med Genet Part A 2011; 155:1661-1667.
(16) Kendall L, Parsons JM, Sloper P, et al. A simple screening method for determining knowledge of the appropriate levels of activity and risk behaviour in young people with congenital cardiac condition. Cardiol Young 2007; 17:151-157.
(17) Rogers P, Mansour D, Mattinson A, et al. A collaborative clinic between contraception and sexual health services and an adult congenital heart disease clinic. J Fam Plann Reprod Health Care 2007; 33:17-21.
(18) Van Damme S, Van Deyk K, Budts W, et al. Patient knowledge of and adherence to oral anticoagulation therapy after mechanical heart-valve replacement for congenital or acquired valve defects. Heart Lung 2011; 40:139-146.
(19) Van Deyk K, Moons P, Farroni N, et al. Effects of a structured education programme for adult patients with congenital heart disease: a follow-up study. Acta Cardiol 2088; 63: 125-126.
(20) Rönning H, Arestedt K, Nielsen NK, et al. Development and Psychometric Evaluation of the Knowledge Scale for Adults with Congenitally Malformed Hearts. J Cardiovasc Nurs 2012; 12: 78-86.
(21) Rönning H, Nielsen NK, Swahn E, et al. Description and initial evaluation of an educational and psychosocial support model for adults with congenitally malformed hearts. Patient Educ Couns 2011; 83:247-251.

(22) Pitcher DW, Papouchado M, Channer KS, et al. Endocarditis prophylaxis: do patients remember advice and know what to do? BMJ 1986; 293:1539-1540.
(23) Goossens E, Fleck D, Canobbio MM, et al. Development of an international research agenda for adult congenital heart disease nursing. Eur J Cardiovasc Nurs 2013; 12:7-16.
(24) Landzberg MJ, Murphy DJ, Davidson WR, et al. Task force 4: Organization of Delivery Systems for Adults with Congenital Heart Disease. J Am Coll Cardiol 2001; 37: 1187-1193.
(25) Deanfield J, Thaulow E, Warnes C, et al. Management of Grown Up Congenital Heart Disease. The Task Force on the Management of Grown Up Congenital Heart Disease of the European Society of Cardiology. Eur Heart J 2003; 24:1035-1084.
(26) Warnes CA, Williams RG, Bashore TM, et al. ACC/AHA 2008 Guidelines for the Management of Adults With Congenital Heart Disease: A report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (Writing Committee to develop Guidelines on the Management of Adults with Congenital Heart Disease). Developed in Collaboration With the American Society of Echocardiography, Heart Rythm Society, International Society for Adult Congenital Heart Disease, Society for Cardiovascular Angiography and Interventions, and Society of thoracic Surgeons. Circulation 2008; 118:714-833.
(27) Day MJ. Educational assessment of the adult with congenital heart disease. Nurs Clin North Am 1994; 29:299-318.
(28) Warnes CA, Liberthson R, Danielson GK, et al. Task force 1: The changing profile of congenital heart disease in adult life. J Am Coll Cardiol 2001; 37:1170-1175.
(29) Yang HL, Chen YC, Wang JK, et al. Measuring knowledge of patients with congenital heart disease and their parents: Validity of the 'Leuven Knowledge Questionnaire for Congenital Heart Disease'. Eur J Cardiovasc Nurs 2012; 11:77-84.
(30) Cohen J. Statistical power analysis for the behavioral sciences. 2nd ed. Hillsdale: New Jersey: Lawrence Erlbaum Associates, Inc., 1988.
(31) Kandula NR, Malli T, Zei CP, et al. Literacy and retention of information after a multimedia diabetes education program and teach-back. J Health Commun 2011; 6:S89-S102.
(32) Roter DL. The outpatient medical encouter and elderly patients. Clin Geriatr Med 2000; 16:95-107.
(33) Schillinger D, Piette J, Grumbach K, et al. Closing the loop: physician communication with diabetic patients who have low health literacy. Arch Intern Med 2003; 163:83-90.
(34) Thompson D, Baranowski T, Buday R. Conceptual model for the design of a serious video game promoting self-management among youth with type 1 diabetes. J Diabetes Sci Technol 2010; 4:744-749.
(35) Baranowski T, Buday R, Thompson DI, et al. Playing for real: video games and stories for health-related behavior change. Am J Prev Med 2008; 34:74-82.
(36) Thompson D, Baranowski T, Buday R, et al. Serious Video Games for Health How Behavioral Science Guided the Development of a Serious Video Game. Simul Gaming 2010; 41:587-606.

9 EFFECTIVENESS OF TRANSITIONAL CARE ON INTERMEDIATE OUTCOMES

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Abstract

Objective: Providing structured patient education is hypothesized to be a critical element of a transition program helping young people with congenital heart disease (CHD) developing the skills, insights, and behaviors needed to transition to adulthood. Evidence supporting the benefit of education on patient knowledge and health-related behaviors is very limited. This study investigated whether a single educational session increased the level of knowledge and changed the prevalence of health risk behaviors in young people with CHD.

Study design: We conducted a longitudinal study of patients transferred to adult CHD care who received one educational session (n=201) at a tertiary care center. Their knowledge level and prevalence of health risk behaviors were assessed using the Leuven Knowledge Questionnaire CHD and the Health Behavior Scale CHD, respectively. A general linear model (GLM) for longitudinal measurements analyzed the natural progression of patients' knowledge over a 27-month period and the effect of one educational session on outcomes.

Results: Participating in an educational session resulted in a small-to-moderate, but significant, increase in total knowledge level and better understanding of deterioration symptoms, and rationale and frequency of follow-up. However, it did not improve patients' health behaviors.

Conclusions: This prospective study is the first to demonstrate the effectiveness of structured education for improving the overall knowledge of young people with CHD. However, this type of education did not improve the patients' tendency to engage in better health behaviors. Urgently needed are studies assessing the effect of repetitive exposure to educational sessions dealing with CHD.

Introduction

Adolescence is a critical and vulnerable period for young people with chronic conditions, such as congenital heart disease (CHD). During this developmental phase, young patients transition to adult life and are expected to develop an increased sense of responsibility by managing their lifestyle, health, and healthcare (1, 2). However, like other teenagers, they are tempted to engage in high-risk health behaviors, such as experimentation with cigarettes, illicit drugs, and binge drinking (3-5). Implementing transition programs is one suggested way to help young people with chronic conditions better navigate the transition into adulthood, teaching them to master a set of new skills, knowledge, and developmental tasks. As young patients grow older, changing healthcare needs demand a change in care setting, that is, from a pediatric- to an adult-focused program. Hence, a timely transfer of care is advocated. Transition programs also can prepare patients for this handing over of care (2, 6-9).

Many authors have proposed definitions for the concepts of transfer and transition (1). Transition can be defined as "*the process by which young people with chronic childhood illnesses are prepared to take charge of their lives and their health in adulthood*" (10). Transfer can be defined as "*an event, or series of events, through which young people with chronic physical and medical conditions move their care from a pediatric to an adult healthcare environment*" (10). Even though the objectives of a transition program for patients with CHD are well documented (2, 11), a clear operational definition for such a program is lacking. Experts have outlined key features and components of CHD transition programs (2, 8, 11, 12). Unfortunately, the critical elements of a formal transition program are not yet clearly defined. Literature promoting the development and implementation of transition programs is extensive (2, 6-9). However, no prospective study has been published on the benefit of such a program in young people with CHD.

Structured patient education has been proposed as a standard element of a transition program (2, 8, 9, 12-14). Developmentally appropriate education about the patients' medical condition and other disease-related issues is hypothesized to be critical in fostering self-management in young people with CHD (2). The objective of structured education is not merely to improve patients' understanding of their disease, but also to encourage patients to adopt a healthy lifestyle, leading to a reduction in morbidity and increase in life expectancy (15-17).

Although patient education is a recommended core component of transition programs, empirical evidence demonstrating its effectiveness in increasing disease-related knowledge in young people with CHD remains scarce (18-20). Furthermore, evidence describing the impact of such

education of young people with CHD on the prevalence of engaging in positive health behaviors is currently absent. Finally, no longitudinal studies have been done or initiated to evaluate the prospective effects of education. Therefore, this longitudinal study had two objectives. First, we sought to describe how disease-related knowledge and engagement of high-risk health behaviors develop in young people with CHD transitioning into adulthood. Second, we sought to determine whether a single structured educational session would increase the level of disease-related knowledge and decrease the prevalence of high-risk health behaviors among these young patients.

Methods

Setting and educational session

This longitudinal study was conducted at a large tertiary care center, housing both a pediatric and an adult congenital heart disease (ACHD) care program. In this center, patients are transferred from pediatric cardiology to ACHD care at the age of 16 years, provided they are medically stable. A multidisciplinary team specialized in adult care provides ACHD care. This team is distinct from the pediatric cardiology team. During a patient's last pediatric visit, the patient is given information on the rationale and timing of transfer to ACHD care. With the consent of all parties, a designated adult provider is generally chosen. This recommendation is documented in the patient's file, and then a pediatric cardiologist writes a referral letter. Currently, a formal educational transition program does not precede this transfer of care within our hospital.

For adults, the frequency of outpatient visits is based primarily on the anatomical classification of the heart defect. The standard frequency of outpatient visits is every 6-12 months for those patients diagnosed with complex heart lesions, every 1-2 years for moderately complex defects, and every 3-5 years for simple lesions (11;13;21). A routine ACHD outpatient visit standardly comprises a consultation with a member of the ACHD advanced practice nursing (APN) team, followed by a medical check-up performed by an ACHD cardiologist. During the APN visit, patients have the opportunity to discuss their health status, symptoms experienced, and pending questions or concerns. Furthermore, every patient receives verbal structured education on disease-related and behavioral issues, including CHD diagnosis; current treatment; rationale for regular follow-up; infective endocarditis symptoms and strategies for preventing it; healthy lifestyle; vocational and educational choices; sexuality; inheritability of the defect; risks associated with the use of contraceptives; and pregnancy. Education and counseling sessions are about 15-30 min. In order to document which items were discussed, repeated, or already known by the patient, the APN team

uses a computerized checklist. Detailed information on the comprehensive list of issues addressed during the education is provided in **Table 9.1.**

Table 9.1: Overview of content and working sequence of structured education session

Congenital Heart Defect
- Description of congenital heart defect
- Anatomical drawing of heart defect on diagram

Treatment regimen
- Medical treatment performed in the past (i.e., surgery, interventional procedures, medication)
- Current medication plan (i.e., name of drugs, dose, indications, special points of attention)
- Future medical treatment (if applicable)

Rationale for regular follow-up
- Rationale for regular continuous cardiac follow-up and requirements (i.e., setting, type of provider, frequency)

Infective endocarditis (IE)
- Definition, characteristics and symptoms of IE
- Preventive measures
- Antibiotic prophylaxis (if applicable)
- Importance of dental hygiene

Healthy lifestyle
- Importance of and requirements of a heart-healthy lifestyle:
  - Cardiovascular risks associated with unhealthy lifestyle
  - Preventive behaviors in terms of substance use, dental hygiene, engagement in physical activity, diet

Vocational and educational choices
- Discussion of choice of education and vocation in adulthood

Sexuality and inheritability
- Sexual concerns relevant for patients with cardiac disease
- Hereditary nature of congenital heart defect
- Information on cardiogenetic services

Pregnancy and contraceptives
- Issues regarding family planning
- Cardiovascular risks associated with pregnancy and delivery
- Choice of contraceptives

Study population

This longitudinal study was conducted as part of the i-DETACH project (Information Technology Devices and Education Program for Transitioning of Adolescents with Congenital Heart Disease). Patients were selected from the database of pediatric and congenital cardiology of the hospital. They were eligible for inclusion if they had a confirmed diagnosis of CHD, were 14-18 years old at the start of the study, had their last outpatient visit at our hospital ≤5 years ago, were able to read and write Dutch, and if their valid contact details were available in the hospital administration. CHD was defined as "*structural abnormalities of the heart and/or great intrathoracic vessels that are actually or potentially of functional significance*" (21). Patients were excluded if they had cognitive and/or physical limitations, preventing them from filling out questionnaires; had undergone heart transplantation; or if they and/or their parents did not consent to participate in the study. Overall, 498 patients met the inclusion criteria.

Over a period spanning three years, four measurements (T_1-T_4) were taken, one every nine months. A set of questionnaires was sent by surface mail to the patients' home address. Patients were asked to fill-out the questionnaires and to return them in a pre-stamped envelope. They received a movie ticket upon completion of the questionnaires. The study was approved by the Institutional Review Board of the University Hospitals Leuven and was performed in line with the principles outlined in the Declaration of Helsinki (22). A total of 429 (Response Rate [RR]=86%), 398 (RR=86%), 366 (RR=82%), and 337 (RR=77%) adolescents participated at T_1, T_2, T_3, and T_4, respectively.

For the present study, data on a selected group of patients were analyzed. Because our aim was to evaluate the impact of a single structured educational session on the knowledge and health risk behaviors of young CHD patients, we analyzed only data from patients who were in current follow-up within our center. Patients who already transferred to the ACHD program prior to the start of the study were excluded from data analysis, because they had already received this type of education. Hence, our final sample comprised 210 patients. Response rates varied between measurement points since a limited number of patients decided to cease participation during the longitudinal data collection (see **Figure 9.1**). Self-reported questionnaires were completed by patients at home and some patients appeared to have missing values for particular items. Complete data for the respective items were available for at least 193 to 196 patients at T1; 183 to 185 patients at T2; 169 to 171

patients at T3; and 149 to 150 patients at T4. These respective ranges varied between measurement points since response rates

Apart from a few exceptions, patients who transferred to ACHD care during the study period had one outpatient visit, and thus were exposed to one educational session. A total of 37, 36, and 33 patients received the educational session after T_1, T_2, and T_3, respectively. One hundred and four patients (49.5%) did not transfer during the study period and therefore were not exposed to patient education (non-education group). **Figure 9.1** presents a detailed flowchart diagram of the sample selection.

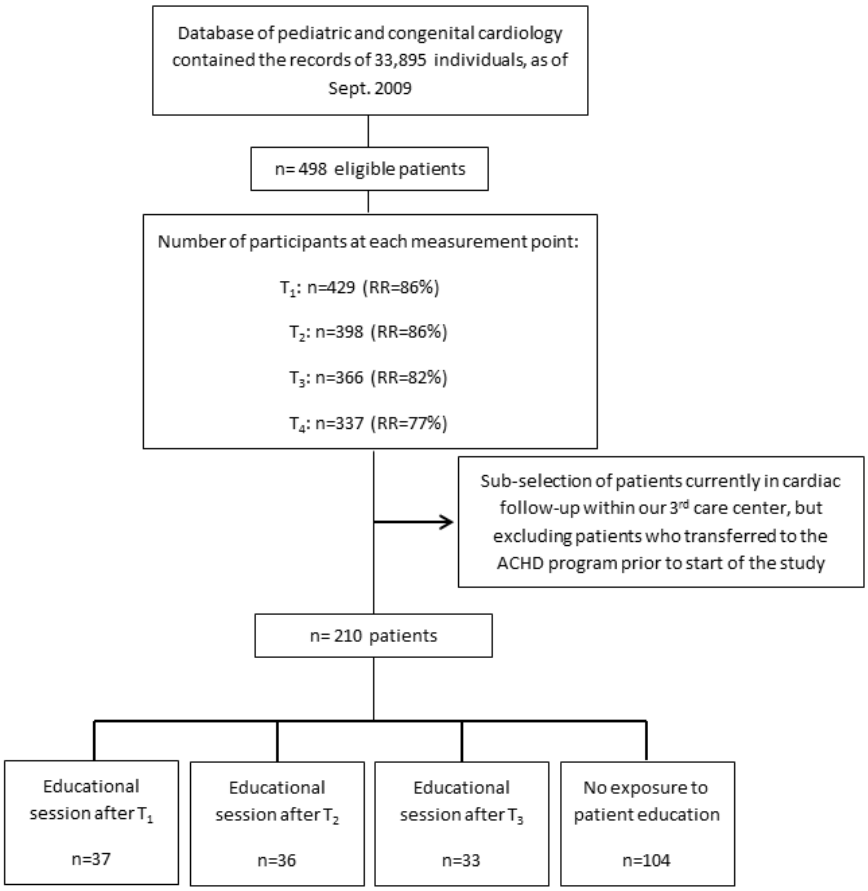
Variables and measurements

Demographic data (i.e., age, sex, and educational level) were collected using a self-report survey. Clinical characteristics were collected by means of chart review and included primary CHD diagnosis, anatomical complexity of the heart defect based on guidance from Task Force 1 of the 32nd Bethesda Conference (13), and history of surgery or catheter-based interventions. The precise date when patients were transferred to ACHD care was determined based on chart reviews. The patient's first ACHD outpatient visit coincides with the first time the patient received structured patient education. The patients were not exposed to this type of education in pediatric cardiology.

The level of disease-related knowledge was measured longitudinally using the Leuven Knowledge Questionnaire for Congenital Heart Disease (LKQ-CHD), which comprises 34 items relevant for female patients and 31 items relevant for male patients (23, 24). For each patient, a total knowledge score was calculated by computing the number of correct answers divided by the number of eligible answers, multiplied by 100. The total knowledge score ranged from 0-100. In addition, eight thematic subscale scores were calculated, representing the level of patient knowledge regarding (1) CHD diagnosis, (2) treatment, (3) follow-up, (4) cardiovascular risk, (5) symptoms, (6) endocarditis, (7) physical activity, and (8) reproduction. These subscale scores also ranged from 0-100 and were calculated as the percentage of correct answers on the respective items comprising these eight subscales. The total knowledge score and the eight thematic scores were calculated for a patient only if at least two-thirds of the survey items were filled out. A patient was said to have adequate understanding if he/she had >80% correct answers, moderate understanding with 50-80% correct answers, and poor understanding with <50% correct answers (24). The LKQ-CHD is a valid instrument for assessing the level of knowledge in patients with CHD and is based on an excellent content validity and relationships with other variables (23).

Health risk behaviors were assessed longitudinally using the 22-item Health Behavior Scale-Congenital Heart Disease (HBS-CHD) (25). This is a self-report instrument that addresses four important components of health behaviors in patients with CHD: (1) the use of alcohol, (2) the use of tobacco and illicit drugs, (3) oral hygiene, and (4) engagement in physical activities. Three health behavior risk scores can be calculated. A substance use risk score (range: 0–3) is based on whether (a) binge drinking occurred at least once a month, (b) ≥ one of seven predefined drugs were used once a month or less, and (c) cigarettes were smoked. A dental hygiene risk score (range: 0–3) is based on whether (a) the patient failed to visit a dentist annually, (b) did not brush his/her teeth daily, and (c) did not floss his/her teeth. Finally, an overall health risk score (range: 0-7) is based on the individual's substance use risk score, dental hygiene risk score, and the absence of sports participation. These three risk scores are transformed into a scale ranging from 0 (no risk) to 100 (maximum risk) in order to facilitate interpretation and to allow for comparisons. The HBS-CHD was found to be a valid and responsive tool for assessing various components of health risk behavior of young people with CHD (25).

Figure 9.1 Flow diagram of subject selection, group comparison, and response rates



Statistical analysis

Differences in demographic and clinical characteristics of patients included in the education group or non-education group were tested using a Chi^2-test for nominal-level data, a Mann-Whitney U test for ordinal-level data, and an unpaired t-test for continuous-level data.

A general linear model (GLM) for longitudinal measurements was used to analyze both the natural progression of patients' knowledge over a 27-month period and the effect of education on LKQ-CHD and HBS-CHD scores. More specifically, a direct likelihood approach was adopted using an unstructured 4x4 covariance matrix for the four longitudinal measurements (26), while considering the measurement point as a categorical predictor (27). The exposure to the educational session was considered to be a binary, non-reversible, time-dependent predictor. That is, the variable is '0' before transfer and '1' after transfer to ACHD care with exposure to structured education. The exact timing of the educational session was determined based on the date at which patients had their first ACHD outpatient visit. For statistical purposes, it was determined whether the educational session was provided to patients after T1, T2 or T3, respectively. At T_1, the baseline levels of knowledge and prevalence of health risk behaviors were determined in 196 patients.

Some basic characteristics of the patients in the present analyses have been previously reported (19, 23, 28). Since significant differences in levels of knowledge according to patients' age; sex; educational level (i.e., high school/college/university, technical high school, vocational high school); and anatomical classification of the heart defect (i.e., mild, moderate, complex) were found (19, 23, 28), these characteristics were added to the model as potential confounding variables here. By including an interaction term between education and when education was provided, the effect of structured education was allowed to differ between T_2, T_3, and T_4. Patients of the same age, sex, education level, and level of CHD anatomical classification were assumed to follow the same knowledge progression as long as they were not exposed to the educational session. This latter assumption allowed for a direct estimation of the effect of the single educational session at the various measurement times and was justified, based on clinical and statistical considerations.

P-values <0.05 were considered to be significant. No corrections for multiple testing were considered.

Sensitivity analysis was also performed using Friedman's repeated measures ANOVA test to assess the robustness of the results obtained through the GLM analysis for the natural progression of

knowledge level and risk behaviors over 27 months. This supplementary sensitivity analysis was performed on the complete data of patients in the non-education group who participated in the four subsequent measurements (n=65). SPSS version 20.0 (SPSS Inc., Chicago, Illinois, USA) and SAS© software, version 9.2 of the SAS© System for Windows (SAS Institute Inc., Cary, NC, USA) were used.

Results

Sample characteristics at baseline (n=210)

Patients had a mean±SD age of 16±1.1 years (range: 14.1-18.2 years). The most commonly diagnosed heart defect was ventricular septal defect (25.7%), followed by aortic valve abnormality (19.5%) and pulmonary valve abnormality (15.7%). Almost half of the sample was diagnosed with a mild defect (49.1%). Moderately and highly complex heart defects were diagnosed in 41.4% and 9.5% of the sample, respectively. More details about demographic and clinical characteristics at baseline (T_1) are presented in **Table 9.2**.

Patients in the education group (n=106) did not differ from patients in the non-education group (n=104) in terms of sex (X^2=.017; *p*=0.897); level of CHD anatomical classification (U=5.708; *p*=0.621); prevalence of cardiac surgery (X^2=.001; *p*=0.970); or educational level (U=4.884; *p*=0.437). Patients of the education group were significantly older than those in the non-education group (16.2±1 years versus 15.7±1 years; t=-3.47; *p*=0.001). However, a difference in mean age of 7 months would likely not be clinically relevant (**Table 9.2**).

Table 9.2: Demographic and Clinical Sample Characteristics at Baseline of Sample (n=210)

Variables		Total sample n=210	Education group n=106	Non-education group n=104	Test statistics
Sex n(%)					
	Male	112 (53.3)	57 (53.8)	55 (52.9)	χ^2= .017
	Female	98 (46.7)	49 (46.2)	49 (47.1)	NS
Age (y) (mean ± SD)		16±1.1	16.2±1	15.7±1.1	t=-3.47 NS
Anatomical classification of primary CHD diagnosis, n(%) [13]					
	Complex	20 (9.5)	12 (11.3)	8 (7.7)	U= 5.708
	Moderate	87 (41.4)	43 (40.6)	44 (42.3)	NS
	Simple	103 (49.1)	51 (48.1)	52 (50)	
Cardiac surgery for CHD, n(%)					
Yes, ≥ 1 cardiac surgical intervention		77 (36.7)	39 (36.8)	38 (36.5)	χ^2= .001
	No	133 (63.3)	67 (63.2)	66 (63.5)	NS
Current level of education, n(%)					
	High school/College/University	90 (46.9)	40 (42.1)	50 (51.5)	U= 4.884
	Technical high school	61 (31.3)	36 (37.9)	25 (25.8)	NS
	Vocational high school	41 (21.4)	19 (20.0)	22 (22.7)	

Baseline assessment of disease-related health-risk behaviors and knowledge level

Before transferring from pediatric to ACHD care (i.e., at T_1) the prevalence of health risk behaviors and the level of knowledge were measured in the overall sample (T_1, n=196). From this baseline assessment, we were able to calculate the health behavior risk scores and the overall and thematic knowledge scores of all participants, both the education group and non-education (control) group.

The prevalence of health-compromising behaviors in the overall sample of patients with CHD (n=196) was low, since the mean±SD overall health risk score was 17.5±14.5 (m_e= 14; IQR= 15), on a scale from 0 to 100. The mean±SD substance use risk score was even lower (5.4±16.7; m_e= 0; IQR= 0). The highest risk score was related to dental hygiene, with a mean±SD risk score of 29.1±24.4 (m_e= 33; IQR= 33).

The overall level of knowledge at baseline was poor, with a total knowledge score of 43±14 for the overall group of patients with CHD (n=210). None of the patients achieved an adequate understanding (>80% correct) on the eight thematic subscales. At baseline, we observed moderate levels of knowledge (50-80% correct) for the subscales CHD treatment (56.2±29.2), rationale and frequency of follow-up (55.6±29.4), and physical activity (67.9±26.2). Poor understanding (<50%

correct) was found for CHD diagnosis (37.2±39.8), endocarditis prevention (40.2 ±22.7), issues related to sexual reproduction (15.7±29.4), cardiovascular risk factors (47.8±20.3), and symptoms of illness deterioration (11.7±32.3).

Impact of structured patient education on health risk behaviors and knowledge level

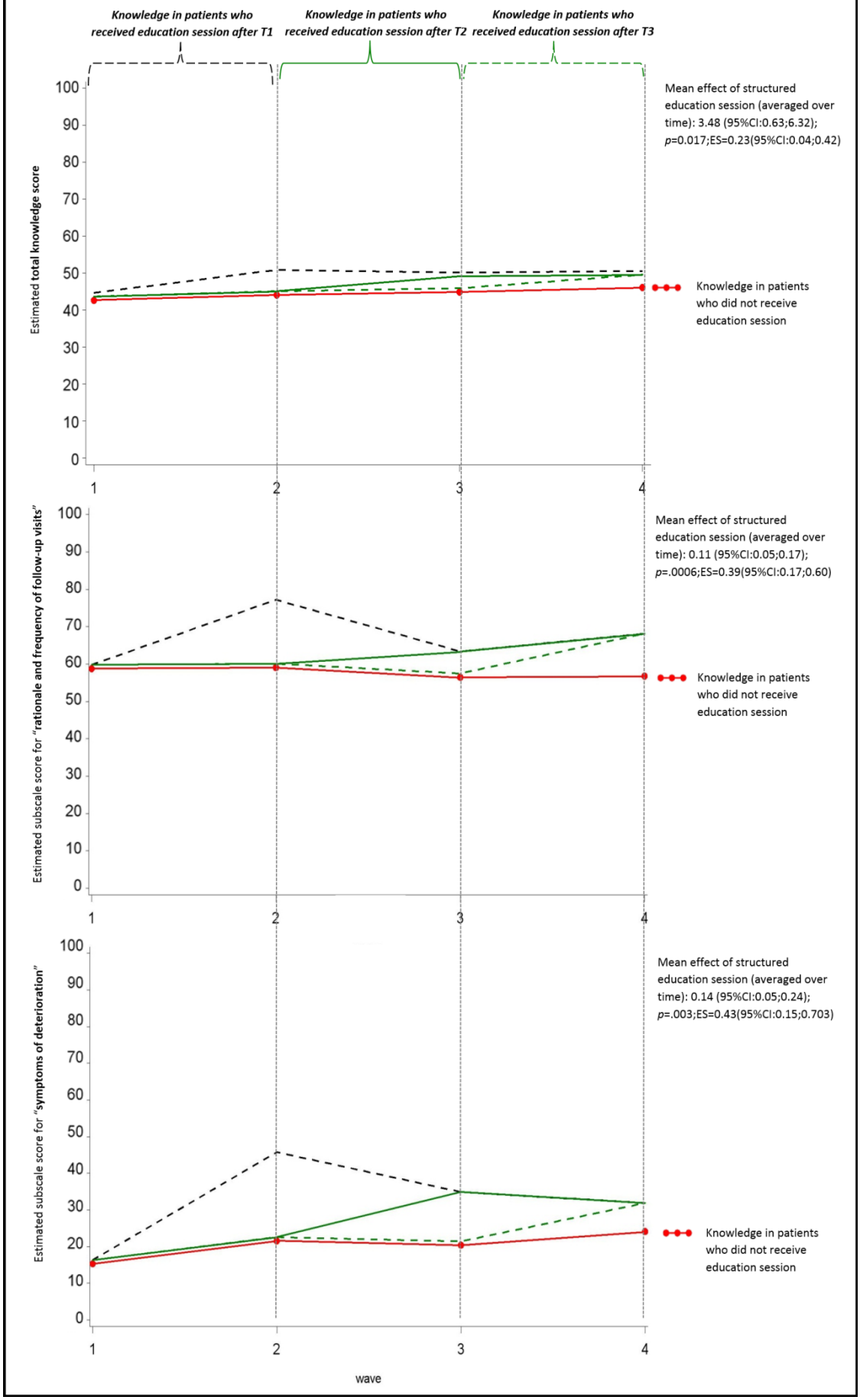
Exactly when patients were exposed to the structured educational session varied. Therefore, we evaluated the effect of education as a function of when it was provided (i.e., after T_1, T_2, or T_3). Because the effect of the educational session might depend on when it was provided during the study, an interaction effect between the timing of education and the actual exposure was tested. This interaction effect, however, was not significant. Hence, the effect of education could be averaged over the respective times of exposure.

Figure 9.2 summarizes the longitudinal evolution of knowledge scores over a period of almost 3 years. Furthermore, this figure illustrates the significant effects of education in four different groups of patients according to the timing at which education was provided. Total knowledge scores increased after exposure to education, regardless of when the session took place (i.e., after T_1, T_2, or T_3 versus non-education group). Providing education produced an average increase in knowledge level of 3.48 (95%CI: 0.63-6.32), and was statistically significant (p=0.017), adjusted for potential confounding factors. The effect size, however, was small (ES=0.23; 95%CI: 0.04-0.42). The effect of education did not depend on when it was provided. That is, the interaction between the timing of the educational session and the actual exposure to the education session was not significant (p=0.726).

Analysis of the eight thematic subscale scores revealed a significant but small effect of education on the subscale that assessed patients' understanding of "symptoms of deterioration" (0.14, 95%CI: 0.05-0.24, p=0.0006; ES=0.39, 95%CI: 0.17-0.60) (**Figure 9.2**). Furthermore, a moderate but significant effect of education was found for the subscale "rationale and frequency of follow-up" (0.11, 95%CI: 0.05-0.17, p=0.0026; ES=0.43, 95%CI: 0.15-0.70) (**Figure 9.2**). Education did not affect the scores of the remaining thematic subscales.

Regarding health risk behaviors, the educational session did not have a significant effect on overall health risk behavior, substance use, and dental hygiene risk scores.

Figure 9.2: Effect of a single education session on knowledge scores (LKQ-CHD) plotted as function of when a session was given



Progression of health risk behaviors and knowledge in patients not exposed to education

The GLM analysis revealed that over time the mean overall knowledge score ($p<0.0001$), overall health risk score ($p<0.0001$), and substance use risk score ($p<0.0001$) increased significantly in patients who did not receive an educational session. The dental hygiene risk score, however, decreased significantly over time ($p<0.0001$) in the non-education group.

These findings were consistent with those from the sensitivity analyses performed on data from a subgroup of 65 patients with CHD who participated in each of the four subsequent measurements and who were not exposed to education during the study. This confirmed that, over time, the prevalence of health risk behaviors in the non-education group increased significantly. Indeed, the mean±SD overall health risk scores (T_1 15.4±13.9 to T_4 30.8±12.7; X^2=60.51; p<0.001) and the mean±SD substance use risk scores (T_1 4.6±15.5 to T_4 15.8±17.7; X^2=35.51; p<0.001) increased from T_1 to T_4 in the non-education group of patients. The dental hygiene risk score, however, did not differ significantly over the study (T_1 25.4±25.7 versus T_4 26.5±25.4; X^2=1.987; p=0.575). The mean total knowledge score increased from 45.6±13.7 at T_1 to 47.9±17.1 at T_4, but this difference was not significant (F=.709; p=0.548) (see T**able 9.3**).

Table 9.3. Results of Sensitivity Analysis: Disease-related Knowledge and Health Risk Behaviors in Patients Who Did Not Participate in Education Session (n=65)

	T_1	T_2	T_3	T_4	Test statistics
Total Knowledge Score (mean±SD)	45.6±13.7	46.3±14.0	47.1±13.6	47.9±17.1	F=.71; p=0.55
Overall Health Risk Score (mean± SD)	15.4±13.9	16.1±14.6	18.1±16.9	30.8±12.7	X^2=60.51; p<0.001
Substance Use Risk Score (mean± SD)	4.6±15.5	5.1±17.9	10.2±22	15.7±17.7	X^2=35.51; p<0.001
Dental Hygiene Risk Score (mean± SD)	25.4±25.7	23.8±21	23.3±25	26.5±25.4	X^2=1.99; p=0.575

Discussion

Within the field of CHD care, there is limited evidence on the benefit of educational interventions designed to improve patients' understanding and knowledge of their condition. Previous studies assessed the effect of education using a matched case-control (19), a pre-post design in a relatively small sample of adults with CHD (18) or a small interventional study without randomization (20). Studies assessing the effect of education on the health risk behaviors of patients with CHD are clearly lacking. To our knowledge, the present study is the first to investigate the prospective effect of education on both knowledge and health risk behaviors in young people with CHD.

In line with previous reports, our study showed that young people with CHD generally possess poor knowledge of their condition prior to education (15, 20, 29-32). Our results also confirm poor-to-moderate levels of understanding of the diagnosis, alarming symptoms, cardiovascular risk factors, reproductive issues, and possible preventive measures against infective endocarditis, as previously reported in a comparable but smaller sample of young people who were not exposed to an educational program (19). Before exposure to the educational session, the prevalence of health-compromising behaviors in our sample was low. This relatively infrequent occurrence of risky behaviors was expected, because published data show low rates of risky health behaviors in comparable samples of young people with CHD (33, 34), and because our respondents were fairly young.

This longitudinal study confirms that one educational session significantly improved overall disease-related knowledge, had a small positive effect on the recognition of symptoms of deterioration, and resulted in a small improvement in knowledge of the rationale and frequency of follow-up visits. During the first ACHD outpatient clinic, the primary aim of the APN team is to introduce young CHD patients to the ACHD team, help them establish a professional relationship with the team, and increase their confidence in the new healthcare team. The education provided at this first meeting predominantly focuses on comprehensively explaining the need for regular cardiac follow-up and how to recognize symptoms requiring medical attention. It is precisely on these subjects that we found a significant effect. Although the educational session appeared to be successful in improving the disease-related knowledge of young patients, the overall effect was small (with a mean increase of 3.5 after exposure to education). This finding emphasizes the need to implement a staged approach when providing education to patients. Gradually building up the content of educational sessions, in combination with a systematic reiteration of important items, is hypothesized to increase retention of information in patients (35-37). Therefore, future longitudinal

studies should assess the potential benefit of consecutive, repeated exposure to educational sessions on the knowledge level of patients in the long term.

Although the primary aim of this educational session is to improve patients' understanding of their disease and treatment, the ultimate goal is to optimize their self-management and behaviors, thereby improving outcomes in the long run. Unfortunately in the present study, a single educational session failed to significantly affect the prevalence of health-compromising behaviors. However, this finding should be evaluated in the context of the pattern of risk behaviors in young CHD patients generally. Longitudinal analysis of the non-education group revealed a significant increase in the overall health risk and substance use scores as they grew older. Hence, young CHD patients tend to acquire more risk behaviors regardless of the type of education, coaching, or guidance they might be exposed to while transitioning to adulthood. One should also bear in mind that the baseline prevalence of risk behaviors in this sample was fairly low, leaving limited room for any possible improvement related to the education. Another possibility is that data were collected for only 3 years, and this might be too brief of a period to detect significant behavioral changes.

Within our hospital, transfer of care to an ACHD program is executed when adolescents with CHD reach the age of 16 years. This transfer is currently not preceded by a formal transition program. However, when reviewing the consensus statements on transition programs, we find that a number of important transition components are in fact currently implemented in our daily practice outside of a formally designed education program. A survey exploring the practices of CHD centers related to transfer and transition showed that for about three-quarters of the centers, the median age of patients at the time of transfer to ACHD care was 18 years. A structured preparation—which participants referred to as a transition program—was only provided in one-third of these transferring centers. The most frequently reported transition-related key elements delivered at the time of transfer was explaining why and how patients would be transferred and providing education about the cardiac condition, treatment plan, and health behaviors (38). At the majority of the centers, these transition-related information sessions took place in both pediatric cardiology and ACHD programs. Only a very limited number of centers had a separate transition clinic in which all these services were provided. In our hospital, pediatric and adult cardiac care is provided in the same building. Patients are prepared for the transfer of care at pediatric cardiology. During their last pediatric visit, the pediatric team discusses the rationale for the transfer, choice of adult provider, and queries the patient about any apprehensions they might have. However, because of logistic considerations and the limited availability of financial and human resources, education, counseling, and guidance relating to adult responsibilities and developmental tasks take place within the ACHD

program after the transfer to ACHD care. When reviewing the interventions performed within our hospital, theoretically, one may conclude that these activities fit within the design parameters of a formal transition program aimed at preparing young people for adulthood.

Methodological considerations

Although a longitudinal observational study design was used, data were collected in a sample of young people receiving care and education within a single tertiary care center, limiting generalizability of our study results. On the other hand, this sample could be considered highly representative of the population of young adults with CHD, because the distribution of CHD anatomical classification levels was in line with those reported in epidemiologic studies (39, 40). Since our ACHD clinic is located in a tertiary care center, which is easily accessible in Belgium, the entire spectrum of CHD is represented at our clinic, including a fairly high proportion of mild CHD. Patients who underwent heart transplantation or patients who had cognitive limitations, however, were excluded from our study. We sent a set of questionnaires to patients four times over a period of three years, and a remarkably high response rate (range: 77%-86%) was achieved, arguing against selection affecting our results. Although data were analyzed for a selection of 210 patients derived from the overall i-DETACH sample (n=429) (25), comparative analyses of the characteristics of these two sample demonstrated a high level of comparability, indicating that selection bias was unlikely. Another possible limitation of the study was that the nine-month intervals between measurement points might have been too brief to detect significant improvements in the respective outcomes, especially in terms of altering health risk behaviors. Furthermore, we can state that during their first outpatient visit at the ACHD clinic, patients were exposed to structured and comprehensive education for the first time. Although we cannot claim that no disease-related information is provided at pediatric cardiology, information is given rather ad hoc than in a systematic and structured way.

Conclusion

This study was the first to demonstrate the prospective effectiveness of structured education in improving the overall understanding of young people with CHD transitioning to adulthood. However, this limited education did not significantly improve patients' health risk behaviors. Additional longer-term studies are needed to assess the effect of repetitive exposure to education on improving knowledge and risk behaviors in young adults with CHD.

References

(1) Meleis AI. Transitions Theory. Middle-Range and Situation-Specific Theories in Nursing Research and Practice. New York: Springer Publishing Company; 2010.

(2) Sable C, Foster E, Uzark K, Bjornsen K, Canobbio MM, Connolly HM, et al. Best Practices in Managing Transition to Adulthood for Adolescents with Congenital Heart Disease: The Transition Process and Medical and Psychosocial Issues: A Scientific Statement From the American Heart Association. Circulation 2011;123:1454-85.

(3) Suris JC, Michaud PA, Akre C, Sawyer SM. Health risk behaviors in adolescents with chronic conditions. Pediatrics 2008 Nov;122:e1113-18.

(4) Staff J, Schulenberg JE, Maslowsky J, Bachman JG, O'Malley PM, Maggs JL, et al. Substance use changes and social role transitions: proximal developmental effects on ongoing trajectories from late adolescence through early adulthood. Dev Psychopathol 2010;22:917 -32.

(5) Chen P, Jacobson KC. Developmental trajectories of substance use from early adolescence to young adulthood: gender and racial/ethnic differences. J Adolesc Health 2012;50:154-63.

(6) American Academy of Pediatrics, American Academy of Family Physicians and American College of Physicians-American Society of Internal Medicine. A consensus statement on health care transitions for young adults with special health care needs. Pediatrics 2002;110:1304-6.

(7) American Academy of Pediatrics, American Academy of Family Physicians, American College of Physicians, Transitions Clinical Report Authoring Group. Supporting the health care transition from adolescence to adulthood in the medical home. Pediatrics 2011;128:182-200.

(8) Foster E, Graham TP, Driscoll DJ, Reid GJ, Reiss JG, Russell IA, et al. Task Force 2: Special health care needs of adults with congenital heart disease. J Am Coll Cardiol 2001;37:1176- 83.

(9) Warnes CA, Williams RG, Bashore TM, Child JS, Connolly HM, Dearani JA, et al. ACC/AHA 2008 Guidelines for the Management of Adults With Congenital Heart Disease: A report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (Writing Committee to develop Guidelines on the Management of Adults with Congenital Heart Disease). Developed in Collaboration With the American Society of Echocardiography, Heart Rythm Society, International Society for Adult Congenital Heart Disease, Society for Cardiovascular Angiography and Interventions, and Society of thoracic Surgeons. Circulation 2008;118:714-833.

(10) Knauth A, Verstappen A, Reiss J, Webb GD. Transition and transfer from pediatric to adult care of the young adult with complex congenital heart disease. Cardiol Clin 2006 Nov;24:619-29.

(11) Rosen DS, Blum RW, Britto M, Sawyer SM, Siegel DM. Transition to adult health care for adolescents and young adults with chronic conditions - Position paper of the Society for Adolescent Medicine. J Adolesc Health 2003 Oct;33:309-11.

(12) Deanfield J, Thaulow E, Warnes C, Webb GD, Kolbel F, Hoffman A, et al. Management of Grown Up Congenital Heart Disease. The Task Force on the Management of Grown Up Congenital Heart Disease of the European Society of Cardiology. Eur Heart J 2003;24:1035-84.

(13) Warnes CA, Liberthson R, Danielson GK, Dore A, Harris L, Hoffman JIE, et al. Task Force 1: The changing profile of congenital heart disease in adult life. J Am Coll Cardiol 2001;37:117-5.

(14) Moons P, Hilderson D, Van Deyk K. Implementation of transition programs can prevent another lost generation of patients with congenital heart disease. Eur J Cardiovasc Nurs 2008 Dec;7:259-63.

(15) Cetta F, Podlecki DC, Bell TJ. Adolescent knowledge of bacterial endocarditis prophylaxis. J Adolesc Health 1993 Nov;14:540-2.

(16) Kantoch MJ, Collins-Nakai RL, Medwid S, Ungstad E, Taylor DA. Adult patients' knowledge about their congenital heart disease. Can J Cardiol 1997 Jul;13:641-5.

(17) Pitcher DW, Papouchado M, Channer KS, James MA. Endocarditis prophylaxis: do patients remember advice and know what to do? BMJ 1986;293:1539-40.

(18) Van Deyk K, Moons P, Farroni N, Dedroog D, Pinxten S, Troost E. Effects of a structured education programme for adult patients with congenital heart disease: a follow-up study. 63 ed. 2008. p. 125 6.

(19) Goossens E, Van Deyk K, Zupancic N, Budts W, Moons P. Effectiveness of structured patient education on the knowledge level of adolescents and adults with congenital heart disease. Eur J Cardiovasc Nurs 2014;13:63-70.

(20) Mackie AS, Islam S, Magill-Evans J, Rankin KN, Robert C, Schuh M, et al. Healthcare transition for youth with heart disease: a clinical trial. Heart 2014;100:1113-8.

(21) Mitchell SC, Korones SB, Berendes HW. Congenital heart disease in 56,109 births. Incidence and natural history. Circulation 1971;43:323-32.

(22) Rickham PP. Human experimentation. Code of ethics of the world medical association. Declaration of Helsinki. 2 ed. 1964. p. 177.

(23) Yang HL, Chen YC, Wang JK, Gau BS, Chen CW, Moons P. Measuring knowledge of patients with congenital heart disease and their parents: Validity of the 'Leuven Knowledge Questionnaire for Congenital Heart Disease'. Eur J Cardiovasc Nurs 2012 Mar;11:77-84.

(24) Moons P, De Volder E, Budts W, De Geest S, Elen J, Waeytens K, et al. What do adult patients with congenital heart disease know about their heart disease, treatment, and prevention of complications? Heart 2001 Jun;86:74-80.

(25) Goossens E, Luyckx K, Mommen N, Gewillig M, Budts W, Zupancic N, et al. Health risk behaviors in adolescents and emerging adults with congenital heart disease: psychometric properties of the Health Behavior Scale-Congenital Heart Disease. Eur J Cardiovasc Nurs 2013;12:544-7.

(26) Molenberghs G, Kenward MG. Incomplete data and SAS. Direct likelihood. Missing data in clinical studies.Hoboken: NJ: John Wiley and Sons; 2007. p. 174-5.

(27) Molenberghs G, Kenward MG. Missing Data in Clinical Studies. Hoboken: NJ: John Wiley and Sons; 2007.

(28) Yang HL, Chen YC, Wang JK, Gau BS, Moons P. An evaluation of disease knowledge in dyads of parents and their adolescent children with congenital heart disease. Eur J Cardiovasc Nurs 2013;28:541-9.

(29) Wang Q, Hay M, Clarke D, Menahem S. Adolescents' drawings of their cardiac abnormality. Cardiol Young 2011;21:556-61.

(30) Van Deyk K, Pelgrims E, Troost E, Goossens E, Budts W, Gewillig M, et al. Adolescents' understanding of their congenital heart disease on transfer to adult-focused care. Am J Cardiol 2010 Dec;106:1803-7.

(31) Kendall L, Parsons JM, Sloper P, Lewin RJ. A simple screening method for determining knowledge of the appropriate levels of activity and risk behaviour in young people with congenital cardiac condition. Cardiol Young 2007 Apr;17:151-7.

(32) Ferencz C, Wiegmann FLJr, Dunning RE. Medical knowledge of young persons with heart disease. Journal of School Health 1980 Mar;50:133-6.

(33) Reid GJ, Webb GD, McCrindle BW, Irvine MJ, Siu SC. Health behaviors among adolescents and young adults with congenital heart disease. Congenit Heart Dis 2008 Jan;3:16-25.

(34) Chen CW, Chen YC, Chen MY, Wang JK, Su WJ, Wang HL. Health-promoting behavior of adolescents with congenital heart disease. J Adolesc Health 2007 Dec;41:602-9.

(35) Kandula NR, Malli T, Zei CP, Larsen E, Baker DW. Literacy and retention of information after a multimedia diabetes education program and teach-back. J Health Commun 2011;16:89-102.

(36) Roter DL. The outpatient medical encouter and elderly patients. Clin Geriatr Med 2000 Feb;16:95-107.

(37) Schillinger D, Piette J, Grumbach K, Wang F, Wilson C, Daher C, et al. Closing the loop: physician communication with diabetic patients who have low health literacy. Arch Intern Med 2003 Jan 13;163:83-90.

(38) Hilderson D, Saidi AS, Van Deyk K, Verstappen A, Kovacs AH, Fernandes SM, et al. Attitude toward and current practice of transfer and transition of adolescents with congenital heart disease in the United States of America and Europe. Pediatr Cardiol 2009 Aug;30:786-93.

(39) van der Bom T, Zomer AC, Zwinderman AH, Meijboom FJ, Bouma BJ, Mulder BJM. The changing epidemiology of congenital heart disease. Nature Reviews Cardiology 2011 Jan;8:50-60.

(40) Hoffman L, Kaplan S, Libertson RR. Prevalence of congenital heart disease. Am Heart J 2004;147:425-39.

10 GENERAL DISCUSSION

This chapter was based in part on: Goossens E., Kovacs A.H., Mackie A.S., Moons P. (2014) Transfer and Transition in Congenital Heart Disease. In: Pediatric and Congenital Cardiology, Cardiac Surgery and Intensive Care. da Cruz E.M., Dunbar I., Jaggers J. (eds.) © Springer-Verlag London 2014

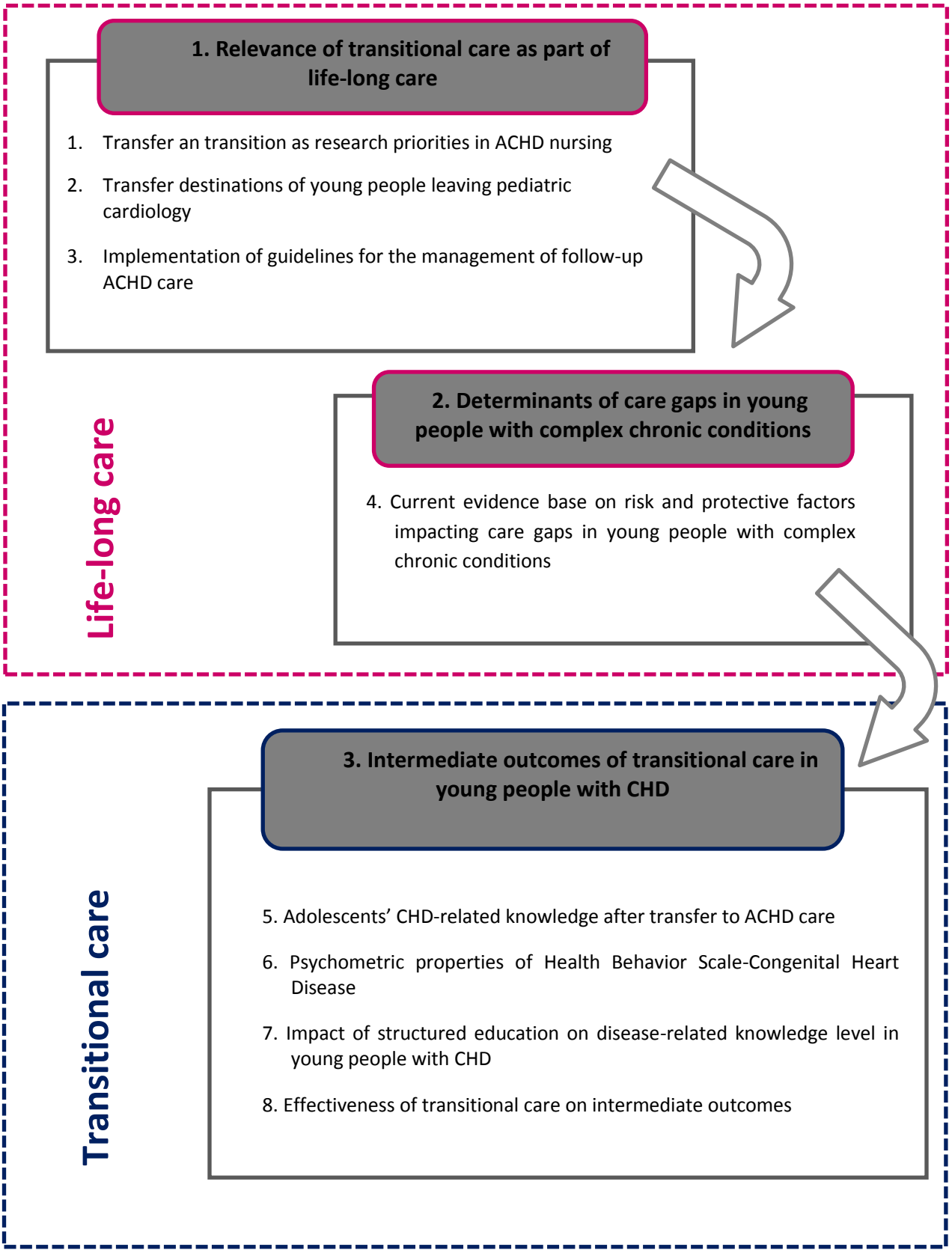
To optimize the life expectancy and quality of life of patients diagnosed with complex chronic conditions, life-long continuity of care is indispensable (1). Congenital heart disease (CHD) is exemplary of such a complex chronic condition. Over the past decades, tremendous investments have been made in the care for patients with CHD through the optimization of CHD detection, performance of early surgery, and long-term management. These efforts were very successful, as they resulted in a substantial increase of the life expectancy. Nowadays, 90% of children born with CHD reach adulthood (2). Despite these improvements, patients with CHD unfortunately have a life-time risk for deterioration of their health condition due to emerging complications. It is estimated that >50% of adults with CHD are at medium-to-high risk for complications such as heart failure, endocarditis, pulmonary hypertension, arrhythmias, and sudden cardiac death (3). Life-long specialized care therefore becomes of paramount importance to safeguard longevity and prevent life-threatening complications (3-5).

International task forces defined the requirements for continuous follow-up care in terms of the frequency of visits and the recommended healthcare setting, based on the complexity of the heart defect and the patient's age (3-5). Indeed, since CHD became a life-cycle disease, patients need to move from pediatric cardiology, over adolescent clinics, to adult-focused settings, and even geriatric care (6). To address the specific care needs of patients at each life stage, age-appropriate care programs are required. During childhood, patients should receive care at specialized pediatric cardiology programs. However, when reaching adulthood, care should be transferred to adult congenital heart disease (ACHD) programs, non-specialist general cardiology care settings, or in shared care programs, according to the heart lesion and healthcare needs of the patient (3-5). This handing-off of care should, however, not occur as an isolated practical event. Since adolescents are experiencing a lot of changes, both from a biological, social, intellectual, and developmental perspective, additional support and counseling should be provided to patients in order to prepare them for the transition to adulthood and the transfer of care. The provision of transitional care, as part of a comprehensive care program, has been proposed as the ultimate strategy to achieve uninterrupted, age- and developmentally appropriate follow-up care throughout the life cycle of patients (7).

This PhD dissertation focused on these two important and highly relevant themes: the importance of life-long care and the provision of transitional care in young people with CHD. Since it remained unclear what the relevance and the magnitude of the problem of gaps in the life-long care process of patients were, this PhD aimed to have a better understanding of this issue by developing an international research agenda for ACHD nursing care (Chapter 2); investigating transfer destinations of young people with CHD in a Belgian setting (Chapter 3); evaluating the level of implementation of ACHD follow-up guidelines in a US setting (Chapter 4); and identifying determinants of care gaps in the overall population of young people diagnosed with a complex chronic condition (Chapter 5). Furthermore, although the importance of developing, implementing, and testing of transitional care interventions in young patients with CHD is well-established, significant gaps remain in our body of knowledge. All consensus documents or recommendations regarding transitional care, stress the importance of an educational curriculum for patients as part of a transition program. Patient education plays a predominant role in transitional care because of its hypothesized value in improving patients' knowledge of their condition and establishment of heart-healthy behaviors (7). Evidence demonstrating the effectiveness of such educational interventions in altering these intermediate outcomes was, however, lacking. This PhD therefore had the second objective to investigate intermediate outcomes of transitional care in young patients with CHD, focusing on disease-related knowledge and health behaviors (Chapters 6-9).

In conclusion, this PhD dissertation is structured around the two general themes of life-long care and transitional care which are addressed in several research questions (**Figure 10.1**).

Figure 10.1: Overview of research objectives addressed in this PhD dissertation



LIFE-LONG CARE FOR PATIENTS WITH CHD

As a result of the markedly improved prognosis of patients born with a congenital heart lesion, CHD became a life-cycle disease. About 90% of patients nowadays have the prospect of achieving adult age (2) and even the population of geriatric patients with CHD is emerging (6). These advances in CHD management, however, bring along new challenges for the current healthcare system. In order to address the age-specific healthcare needs of patients with CHD, age- and developmentally care programs are mandatory at every stage of life. Since patients with CHD are affected at birth, their care process generally starts at pediatric cardiology. As patients grow older, a shift of care from pediatric cardiology, over adolescent care to adult, and even geriatric care is required. Understandably, such complex transfers of care across settings make patients vulnerable for discontinuation of their care process. Such discontinuations leading to exceeding the recommended guideline-based period of time between mandatory follow-up visits are called 'care gaps'(8).

Previous studies indicated that such care gaps occur regularly in several types of complex chronic conditions (9-13). Furthermore, the period during which patients developmentally transition to adulthood seemed to be characterized by an increased vulnerability for this care problem. Empirical evidence demonstrated that gaps in the transitional care process are observed in about a quarter of young people with acromegaly (9;10;14), Human Immunodeficiency Virus (HIV) -infection (15), or diabetes mellitus type I (16). When focusing on young people with CHD, the problem of care gaps, however, appears to be most prominent in this population (17-22). Studies reported that 21 to 76% of adolescents with CHD experienced care gaps, or are even completely lost to follow-up (17-22).

Empirical evidence demonstrated that care gaps are present in a noteworthy proportion of young patients with a complex chronic condition, such as acromegaly (9), diabetes mellitus type I (16), and juvenile idiopathic arthritis (23). The occurrence of care gaps seemed, somewhat surprisingly, to be the highest in patients with CHD. Systematic literature reviews on transfer and transition in chronic conditions, however, revealed that this issue is currently predominantly studied in patients with CHD (24-27). Due to a lack of studies investigating the problem of care gaps in other populations of young people with a chronic condition, it remains currently unknown whether this problem is truly more prominent in patients with CHD as compared to other conditions, or seems to be of significance due to the fact that it is currently more frequently studied in patients with CHD (i.e., the numbers tell the tale). Furthermore, based on the results derived from the systematic review of literature describing risk factors for care gaps in patients with complex chronic conditions,

an important number of identified risk factors are frequently present in patients with CHD. For example, a significant proportion of patients has a mild heart lesion, do not need to undergo surgical interventions, encounters healthcare insurance problems, or need to travel a large distance in order to reach the nearest specialized adult clinic. These respective patient characteristics were found to increase the likelihood of experiencing care gaps. Additional studies are, however, needed in order to support this hypothesis.

This growing interest in studies investigating transfer and transition was also reflected in the list of research priorities established as part of this PhD (Chapter 2). Based on the opinion of about 40 nurse specialists and researchers within the field of ACHD nursing, we learned that studies investigating transfer and transition are one of the top 5 priorities for future research (28). Although this mixed methods study was the first to develop a research agenda for ACHD from an international perspective, this study had a rather limited scope since the expert panel consisted only of nurse specialists and researchers. Ideally multiple perspectives, including the opinion of CHD clinicians, patients, families, and policy-makers should be investigated in order to develop a comprehensive research agenda which could direct future research initiatives. After the publication of our list of research priorities for ACHD nursing, two additional publications reported on the development of a science agenda for CHD (29;30). Based on the report from the Centers for Disease Control and Prevention (CDC) Experts Meeting (2013) we can conclude that priority should indeed be given to studies focusing on the continuation of care from adolescence to adulthood in patients with CHD. This CDC initiative developed a research agenda based on the perspectives of medical doctors, surgeons, epidemiologists, public health officers, patient advocates, and patients themselves (29). The list of clinical research priorities that was developed by the Alliance for Adult Research in Congenital Cardiology (AARCC), however, did not include any research priorities related to transfer, transition, or the challenges of providing life-long care (30). This study, nonetheless, aimed to exclusively identify clinical research objectives related to treatment, surgery, and medical management of CHD.

Levels of care

Since data were lacking on the level of care continuity of young people with CHD within the Belgian healthcare system, we investigated the destinations of transfer of patients known to be cared for at pediatric cardiology at the University Hospitals Leuven (Chapter 3) (18). This study revealed that only a small proportion of patients ceased cardiac follow-up (6.2%) or appeared to be untraceable within the healthcare system (1.1%). The majority of patients (80%), however, remained at the specialized cardiac care level. Only a minority (2.2%) of young people with CHD apparently did not transfer to adult care and thus remained within the pediatric program. When comparing the results obtained in this single center Belgian study with published literature, it was clearly noticed that the proportion of patients experiencing a care gap was significantly lower as compared to the 21-76% reported previously (17;21;22;31-33). Since comparability between studies is, however, rather limited due to important differences in study methodology, we replicated this initial study in a sample of young adults with CHD known to be cared for at the Children's Hospital Boston, USA (Chapter 4). Using the same research methodology, the implementation of recommendations regarding the level of ACHD care was evaluated in this sample. As part of this study, the setting of care, frequency of outpatient visits, and healthcare professional providing follow-up care were evaluated. Despite the uniform study methodology, important differences were found. While the proportion of patients who ceased cardiac follow-up was almost comparable (6.2% versus 6.5%), other important differences appeared.

First, though in both samples the majority of young adults received care within a specialized cardiology setting, the proportion of patients not having made the transfer to ACHD care was surprisingly larger in the Boston sample (48%) as compared to the Leuven sample (2%). Within the University Hospitals Leuven, an internal policy is implemented resulting in the transfer of patients to ACHD care around the age of 16 years. Furthermore, from a practical point of view, patients do not necessarily have to change their care to another hospital since pediatric cardiology and the ACHD care program are located under the same roof. In order to facilitate the transfer of care, clinical information of patients is shared between both care programs through the use of an electronic database. It can be hypothesized that these organizational components (i.e., transfer policy, location of programs, and shared electronic database) might support the transfer of care to ACHD, although empirical evidence supporting this hypothesis is currently lacking.

Second, this contrast in the proportion of patients remaining in pediatric cardiology might be related to significant differences in the availability of ACHD specialists across areas and regions

hampering transfer of care to ACHD care. Belgium is a fairly small country with a large population density, in which the travel distance for patients to arrive at the closest ACHD center did not emerge as a determinant of cessation of cardiac care (18). Although, to date two additional studies performed in adolescents with CHD reported that the travel distance to the closest ACHD center was not a determinant of care gaps, this variable was a significant risk factor in patients with sickle cell disease (11) and congenital adrenal hyperplasia (23).

Currently, a limited number of studies explored barriers to transferring young patients towards ACHD care (8;33-35). Besides institutional factors such as unlimited age access to adult care or a limited capacity of ACHD centers (34), mainly patient-related barriers were reported. Examples of these latter barriers are: non-compliance with the transfer plan, emotional or cognitive delay, health insurance issues or unawareness of need for ACHD care (8;33;35). Although one could argue that the recommendation, stating that a transfer of care towards adult-focused care is mandatory, is based on expert opinion, preliminary evidence demonstrated the benefit of transferring patients to ACHD care (36). Mylotte and colleagues was the first to demonstrate a significant association between decreased mortality rates and increased referral rates towards ACHD care following the publication of national consensus guidelines on ACHD care in Canada.

Untraceability

Although the proportion of patients self-reporting cessation of cardiac follow-up was comparable between the Leuven (6.2%) and Boston (6.5%) samples, a large contrast was, however, observed with regard to the number of patients who were untraceable in the healthcare system (1.1% versus 24.9%, respectively). To our opinion, losing sight of patients due to their untraceability within the healthcare system is very closely related to becoming entirely lost to follow-up. To date, with the exclusion of this PhD study, only two additional studies explicitly investigated the problem of untraceability of young patients with CHD (18;34). In these latter studies, untraceability was an issue in a small (1.1%) (18), moderate (12%) (34), and large proportion of young patients with CHD (24.9%). The difference in prevalence rates of untraceability between these samples indicates that multiple factors including characteristics of geography, population, and healthcare systems might play a prominent role in this issue. Our study was, however, the first to explore potential determinants of untraceability (Chapter 4). Patients who underwent none or a lower number of heart surgeries in the past; had health insurance issues; or were of a non-white ethnicity had an increased risk for becoming untraceable. Furthermore, it could be hypothesized that differences in migration of patients between countries or states might play a role. Indeed, US-citizens are known

to move more often in-between states or countries as compared to Belgian citizens. This difference can be demonstrated when emigration rates from that state of Massachusetts (2.4% in 2012) (38) is compared to Belgium (0.8% in 2010) (39). Hence, studies that provide empirical evidence describing the magnitude of this problem and identifying potential drivers of untraceability are highly needed. The development and implementation of (inter)national databases which keep track of patients with CHD throughout their life and within the overall healthcare system, might be one strategy to tackle the problem of untraceability (8;29;40).

Specialized follow-up care

Though it might be encouraging to observe that about three quarters of young patients with CHD remain in specialized follow-up care, one might question if all these patients actually need to be cared for at this highly specialized level. Indeed, based on the hierarchical algorithm for ACHD care provision, only a selected group of patients diagnosed with a highly complex heart defect is recommended to receive care exclusively provided by ACHD specialists (3-5;41;42). Patients with moderately complex CHD characterized by an uncomplicated disease course could, however, also be cared for within shared care facility. Shared care is the level of follow-up in which care is provided by community providers and ACHD specialist collaboratively. Such shared care could, however, be predominantly organized in regional hospitals, dispersing care across the larger healthcare system. Based on the insights gained in this PhD, unfortunately, we have to conclude that shared care is only provided in a limited number of cases. Future efforts should be made to establish shared care networks in a formal way, facilitating the transfer of selected patients to this level of care. Hence, in order to safeguard the accessibility and affordability of the current healthcare system, strategies should be developed. Since the group of adults with CHD will likely grow exponentially during the upcoming decades, existing ACHD care centers might become saturated and problems like waiting lists might become inevitable. One of the proposed strategies, which might safeguard the current easy accessibility of tertiary care, is the implementation of the principles of subsidiarity. Subsidiarity can be defined as the provision of care at the least complex level that is clinically appropriate to the patient's condition (43). In general, lower levels of care are associated with lower healthcare costs and with less fragmented, but more continuous and coordinated care (44). Over the past decades, the provision of care for a number of chronic conditions such as diabetes mellitus and chronic renal insufficiency was therefore successfully shifted from secondary (i.e., specialized) toward primary levels (45-47). Since an important proportion of patients with CHD are medically stable during specific life stages and can function relatively well without continuous surveillance in tertiary care centers, this principle of subsidiarity might be of value.

Currently the delivery of care across different levels of care is grounded on expert-based recommendations. Unfortunately, the evidence base underpinning these consensus documents is poor with less than 1% of recommendations grounded on level A-evidence (3). This hierarchical algorithm for follow-up care determines the most appropriate level of care according to patient-related characteristics such as: medical stability, CHD complexity level, and potential risk for long-term complications. One could, however, argue that a list of additional criteria such as predicted clinical outcomes and the costs of care are more appropriate, valid, and relevant as a basis to choose the level of care. Unfortunately, empirical data underpinning such a strategy for redirection of care to lower levels is non-existing at the moment. Hence, there is a need for the development and investigation of an empirically-based algorithm for the assignment of patients with CHD to different levels of care, in accordance with the principles of subsidiarity. Such an algorithm can guide clinicians and policymakers to refer patients to the most optimal level of care by obtaining the best clinical outcomes at the fairest price. Such efforts are of paramount importance to make CHD care sustainable and affordable within the context of the numerous challenges that our current healthcare system will most likely face in the future.

Determinants of care gaps

Tailored interventions preventing care gaps should focus on identified risk factors. Based on a systematic literature review performed as part of this PhD, we learned that nowadays a limited set of patient-related characteristics is known to influence the risk of experiencing care gaps (Chapter 5). Four categories of patient-related characteristics were identified in young people afflicted with complex chronic conditions: demographics, disease-related characteristics, healthcare services use, and patient's health behaviors and beliefs. Although most identified risk factors are only modifiable to a limited extent, some protective factors such as guaranteeing that patients leave pediatric care with a written recommendation on the type of professional providing follow-up adult care, or checking if patients attend the first outpatient visits in adult care, might be components of interventions preventing care gaps. Although this systematic review provided a set of 19 determinants of care gaps, all factors were identified on the patient level. Comparison of the results of studies currently performed in patients with CHD, however, revealed substantial differences between countries or healthcare systems with regard to the proportion of patients experiencing care gaps. Such differences cannot be explained solely on the basis of the known patient-related determinants of care gaps. This observation formed the basis for the development of a hypothesis stating that determinants of care gaps most likely have a multi-factorial and multi-level nature. In addition to patient-related factors, aspects of care providers, the organization of care within the

institution, and the characteristics of the healthcare system are believed to have an important impact on continuity of care (18;20). Healthcare system and organizational factors, such as the introduction of co-payment and obligatory transfer policies, have yet to be systematically evaluated. In order to scrutinize this hypothesis, an international, multicentric, multi-level study investigating healthcare system, organizational and patient-related characteristics associated with care gaps in patients with CHD is imperative.

Based on the results obtained as part of this PhD, it became clear that the occurrence of gaps in the care process is an important problem during the adolescence and young adulthood, although the prevalence differs across healthcare systems (Chapters 3 & 4). To date, it is, however, unknown to what extent care gaps occur during other life stages. Furthermore, the clinical impact of such care gaps is poorly understood. In patients with CHD, such care gaps might potentially aggravate disease burden, as the complex nature of the heart defect and its inherent high risk for comorbidity onset warrants close and sustained follow-up. Preliminary results indicated that care gaps in patients with CHD are associated with an increased risk for developing comorbidities and a need for urgent interventions (17;32), although evidence demonstrating the effect of care gaps in patients with CHD is currently scarce.

TRANSITIONAL CARE

During the developmental transition towards adulthood, young people with CHD are supposed to attain a set of skills, insights, attitudes, and behaviors deemed mandatory in order to function autonomously within adult life and the adult healthcare system. In order to achieve this set of developmental tasks, adolescents should be supported and challenged to accomplish skills related to decision-making, self-advocacy, and self-efficacy (48). The provision of transitional care is suggested as the most relevant strategy to accomplish these goals (7). Based on the available consensus statements regarding transitional care for young people with CHD, an educational curriculum is considered to be an indispensable component of a transitional care program. The evidence base supporting this recommendation and demonstrating the beneficial effects of providing education is, however, very poor. Patient education is hypothesized to increase patients' knowledge of their condition, treatment regimen, preventive measures, and lifestyle matters (7). Furthermore, it is assumed that education will enable patients to gradually assume all responsibilities related to adult life, health, and healthcare. Ultimately, education is provided with the initial objective to increase patients' knowledge, but secondarily aims to result in the establishment of heart-healthy behaviors. This PhD was the first to address significant gaps in the knowledge base regarding the effectiveness of educational interventions in young people with CHD as part of transitional care provision.

Based on observations made in adults with CHD, demonstrating a poor level of disease-related knowledge (49-52), it could be hypothesized that adolescents and young adults most likely have significant shortcomings in their knowledge level. This PhD was the very first to investigate the level of knowledge in adolescents at several moments in their transitional care process (i.e., pre- and post- transfer to ACHD care) (53;54). As anticipated, significant gaps in the level of knowledge were identified in this population (Chapter 6). Although patients demonstrated adequate to moderate levels of knowledge on a limited set of CHD-related topics, poor knowledge was observed for most prominent topics such as the specifications of the defect, risk and recognition of life-threatening complications, and healthy lifestyle matters (54;55).

These findings demonstrated there is significant room for improvement and a potential pathway for educational interventions to improve the level of disease-related knowledge. Several studies performed as part of this PhD, enlarged the evidence base regarding the effectiveness of educational interventions on two intermediate outcomes, disease-related knowledge and health risk behaviors (Chapters 8 & 9).

Disease-related knowledge

Using several samples, research designs, and statistical analysis techniques, this PhD demonstrated that the provision of structured CHD education was an independent determinant of higher levels of knowledge in young people with CHD, irrespective of the age, educational level or complexity of the heart defect (Chapter 8). Furthermore, longitudinal analyses confirmed that one educational session significantly improved overall disease-related knowledge, had a small positive effect on the recognition of symptoms of deterioration, and resulted in a small improvement in the level of patients' knowledge regarding the rationale and frequency of follow-up visits. Despite the fact that the provision of one educational session proved to significantly improve the level of knowledge, this effect was rather small (Chapter 9).

Furthermore, although the provision of education proved to be an independent determinant of higher levels of knowledge, patients with more complex levels of CHD were found to have significantly higher levels of knowledge. Since patients with highly complex heart defects are more frequently exposed to education during the yearly outpatient visits, one could hypothesize that repeated exposure to education might result in a more prominent improvement of knowledge. Unfortunately, this hypothesis remains currently unstudied since no study in patients with CHD assessed the effect of an intervention comprising multiple consecutive education sessions on the knowledge level of individual patients. Future studies should investigate the potential dose-effect relationship between the number of education sessions and the total knowledge score of patients.

As stated before, the magnitude of the effect gained through the provision of one educational session was rather disappointing. Despite the fact that significant resources (e.g., time, personnel) were invested, a small mean increase of 3.5 (scale 0-100) was obtained after exposure to the educational intervention. Other alternative interventions, methods, and strategies aiming to improve the disease-related knowledge of patients could, however, have a larger effect. Examples of such interventions are the provision of written information, organization of group education sessions, use of computerized education programs (56), or application of motivational interviewing techniques. On the other hand, since evidence is lacking on the potential benefit of repeated exposure to education, it currently remains unknown whether a large effect can be obtained by repeating this educational sessions over a period of time in individual patients.

Health behaviors

With regard to altering the health behaviors of adolescents with CHD, providing one single session of education did not appear to be an effective strategy since our longitudinal study could not demonstrate a significant improvement of the health behaviors of patients. Within our sample of more than 400 adolescents, risky health behaviors were relatively rare. Previous studies, conversely, reported that adolescents are more likely to experiment with alcohol, illicit drugs, and risky sexual behaviors (57-60). In order to enlarge the generalizability of our study findings, additional studies are required. Both studies describing the health behaviors of young people with CHD in a comprehensive way, as well as studies scrutinizing the effect of interventions such as patient education or the motivational interviewing technique on the lifestyle of patients are mandatory.

As part of this PhD, the Health Behavior Scale – Congenital Heart Disease (HBS-CHD) was developed as a comprehensive instrument for the assessment of self-reported health behaviors of patients with CHD (Chapter 7). In order to investigate the psychometric properties of this instrument, the HBS-CHD was completed four times over a period of three years in a sample of about than 400 young people with CHD and 400 peers from the general population. Based on the results of this study, the HBS-CHD was found to be valid and responsive questionnaire to assess health risk behaviors in young people with CHD. Future studies are, however, needed in order to test the stability of the instrument and to provide evidence support its psychometric properties in other age groups such as adults with CHD.

IMPLICATIONS FOR PRACTICE

This PhD substantially enlarged the evidence base indicating that transition towards adulthood is a vulnerable period for young patients with CHD. Problems such as the occurrence of care gaps, cessation of cardiac follow-up, or patients becoming untraceable in the healthcare system were identified in diverse samples of young people across several healthcare systems. Furthermore, young people's vulnerability was also demonstrated by the gaps in their disease-related knowledge and the slight increase of health risk behaviors around the age of transfer to ACHD care. These findings indicated there is a clear need for interventions, strategies and transitional care programs tackling these problems encountered in daily practice.

Life-long care for patients with CHD

Based on the results of this PhD, preliminary data are provided which could help healthcare professionals to identify patients who have an increased risk for either receiving care that is not totally in line with the current set of recommendations; at risk for stopping cardiac follow-up care; or at risk for becoming untraceable in the healthcare system. Based on this set of determinants, healthcare workers could screen for risk populations.

Although the evidence base identifying robust, valid, and relevant determinants of care gaps is relatively poor, healthcare professionals could implement components protective for care gaps such as providing written recommendations on the requirements for adult follow-up in patients' medical file; or tracking the attendance rates of patients at their first outpatient visits in the adult clinic.

Furthermore, a registry for patients with CHD throughout their entire life spectrum might be of value both from a clinical as well as a scientific point of view. Keeping track of patients from birth until death, enables healthcare providers to comprehensively follow the progress of the heart condition throughout life. Furthermore, such a registry could at an early stage identify patients who have missed appointments, are non-compliant to the proposed follow-up plan, experience care gaps or became untraceable in the healthcare system.

The establishment of care networks in which care for patients with CDH is shared between specialist and non-specialist cardiologists should be explored. Redirection of patients to lower levels of care is deemed mandatory to safeguard the easy accessibility of tertiary care in the upcoming decades. This PhD concluded that shared care facilities are currently only used in a limited number of

patients. To advantages of redirecting patient care to shared care facilities in terms of preserving easy accessibility of tertiary care for patients with complex CHD, clinical outcomes, and associated healthcare expenditures should, however, be scrutinized.

Transitional care

Based on the current recommendations regarding transitional care, education and counseling are hypothesized to be highly important components of a transition program for adolescents with CHD. Data clearly demonstrated significant gaps in the knowledge of patient regarding their condition, treatment, preventive measures, and implications of the condition in adult life. The provision of structured, tailored education during a face-to-face consultation with adolescents results in a significant although small improvement of the disease-related knowledge. No improvement of health behaviors can, however, be expected after providing one educational session. Innovative supplementary interventions aiming to increase patients' level of disease-related knowledge must be developed and tested in practice.

The prevalence of health-compromising behaviors appeared to be fairly low in our sample of adolescents with CHD. These data, however, clearly demonstrated a significant increase of experimenting behaviors around the age at which patients generally transfer to ACHD care (i.e., 16 years). This finding indicated a need for actively detecting risky behaviors in patients. This can be done in a valid way by using the HBS-CHD. Altering the health behaviors of young people through the provision of education seemed, however, challenging and rather ineffective.

AVENUES FOR FUTURE RESEARCH

Although this PhD delivered an important contribution to the body of knowledge regarding transitional care for young people with CHD, significant gaps remain in our understanding of this important issue. Several avenues for future research are proposed.

Life-long care for patients with CHD

Firstly, a large variability in prevalence rates of care gaps across previous studies could be observed. It is unknown whether these differences originate from different study methodologies (e.g., used definitions, inclusion criteria, and recruitment settings) or reflect true differences between patients, hospitals, and/or healthcare systems. Hence, an international study, across different healthcare systems, exploring the predictive value of multi-level determinants of care gaps is required. In order to prevent such gaps in the care process of patients, tailored interventions need to be developed, implemented, and tested in practice. The preliminary set of determinants of care gaps identified through our systematic literature review in young people with complex chronic conditions, could provide evidence-based components to be included in such interventions. The number of studies exploring potential determinants of care gaps is, however, limited at the moment. Additional studies could add to our current understanding of this issue.

Second, previous studies focused on the prevalence and determinants of care gaps during the developmental transition of young people with CHD towards adulthood. However, since CHD became a life-cycle disease, patients will have to transfer care across several settings at multiple life stages. Therefore, studies scrutinizing care gaps along the entire life cycle of patients with CHD, from birth to death, are highly needed. Moreover, although life-long care is recommended by experts to prevent serious complications (3;5), evidence demonstrating the impact of care gaps on clinical outcomes is very limited (17;32). One study on about 150 adults with moderate-to-complex CHD living in the Unites States revealed that 60% of patients with a care gap were diagnosed with a secondary cardiac abnormality. Care gaps were furthermore associated with a threefold greater need for urgent cardiac interventions (17). A large scale assessment of the impact of care gaps in terms of mortality, morbidity, and healthcare services use is, however, missing.

Besides the fairly well described problem of care gaps occurring during the transition of patients towards adulthood, this PhD identified important differences in the number of patients appearing untraceable across different healthcare systems. Since the true magnitude of this problem is poorly understood, additional studies determining the number of untraceable patients and

identifying determinants are needed. In order to guide such research initiatives, additional initiatives should be undertaken in order to provide a clear conceptual and operational basis for the concept of untraceability. There is currently a lack of consensus regarding the difference between patients who ceased cardiac follow-up, are classified as being lost to follow-up, or are identified to be untraceable.

Finally this PhD reported that a significant number of young adults with CHD are cared for in highly specialized ACHD centers. Since the number of adult patients is expected to grow exponentially in the upcoming decades, strategies must be developed to safeguard the easy accessibility of tertiary care to patients with highly complex CHD in the first place. Future studies should explore the potential benefits of implementing the principle of subsidiarity in clinical practice. Outcomes of assigning patients to less specialized levels of care, in terms of mortality, morbidity, healthcare services use, and healthcare expenditures should be investigated thoroughly. Furthermore, there is a need for the development and investigation of an empirically-based algorithm for the assignment of adults with CHD towards the most appropriate and cost-effective levels of care.

Transitional care

In order to strengthen the generalizability of our findings demonstrating significant shortcomings in the levels of disease-related knowledge of adolescents with CHD, complementary studies are needed. As reported in our mixed methods study establishing an international research agenda for ACHD nursing, there was a vast consensus that the highest priority in future studies should be given to studies describing the level of knowledge and assessing the benefit of providing patient education. Identified research priorities related to knowledge and education of patients with CHD comprised the description of the disease-related knowledge of patients; the development of an educational plan for life; and having special attention for patients dealing with learning difficulties or neuro-cognitive impairments.

Although a significant but small effect of patient education was demonstrated in this PhD, several questions remain unanswered. Future studies comparing the effectiveness and effect size of several educational interventions such as a series of consecutive educational sessions, the provision of written information pamphlets, the use of computerized education programs or the implementation of the motivational interviewing technique need to be performed.

Finally, there is a need for studies evaluating the psychometric properties of the HBS-CHD in adult populations derived in different centers, countries, continents, and even cultures. Such studies

should evaluate the validity, relevance and responsiveness of this instrument worldwide in different samples of patients with CHD.

References

(1) Chen LM, Ayanian LZ. Care continuity and care coordination: what counts? JAMA Intern Med 2014;174(5):749-750.

(2) Moons P, Bovijn L, Budts W, Belmans A, Gewillig M. Temporal Trends in Survival to Adulthood Among Patients Born With Congenital Heart Disease From 1970 to 1992 in Belgium. Circulation 2010;122(22):2264-2272.

(3) Warnes CA, Williams RG, Bashore TM, Child JS, Connolly HM, Dearani JA et al. ACC/AHA 2008 Guidelines for the Management of Adults With Congenital Heart Disease: A report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (Writing Committee to develop Guidelines on the Management of Adults with Congenital Heart Disease). Developed in Collaboration With the American Society of Echocardiography, Heart Rythm Society, International Society for Adult Congenital Heart Disease, Society for Cardiovascular Angiography and Interventions, and Society of thoracic Surgeons. Circulation 2008;118(23):714-833.

(4) Deanfield J, Thaulow E, Warnes C, Webb GD, Kolbel F, Hoffman A et al. Management of Grown Up Congenital Heart Disease. The Task Force on the Management of Grown Up Congenital Heart Disease of the European Society of Cardiology. Eur Heart J 2003;24:1035-1084.

(5) Baumgartner H, Budts W, Chessa M, Deanfield JE, Eicken A, Holm J et al. Recommendations for organization of care for adults with congenital heart disease and for training in the subspeciality of 'Grown-up Congenital Heart Disease' in Europe: a position paper of the working Group on Grown-up Congenital Heart Disease of the European Society of Cardiology. Eur Heart J 2014;35(11):686-690.

(6) Baumgartner H. Geriatric congenital heart disease: a new challenge in the care of adults with congenital heart disease. Eur Heart J 2014;35:683-685.

(7) Sable C, Foster E, Uzark K, Bjornsen K, Canobbio MM, Connolly HM et al. Best Practices in Managing Transition to Adulthood for Adolescents with Congenital Heart Disease: The Transition Process and Medical and Psychosocial Issues: A Scientific Statement From the American Heart Association. Circulation 2011;123:1454-1485.

(8) Gurvitz M, Valente AM, Borberg C, Cook S, Stout K, Kay J et al. Prevalence and Predictors of Gaps in Care Among Adult Congenital Heart Disease Patients (The Health, Education and Access Research Trial: HEART-ACHD). J Am Coll Cardiol 2013;61(21):2180-2184.

(9) Delemer B, Chanson Ph, Foubert L, Borson-Chazot F, Chabre O, Tabarin A et al. Patients lost to follow-up in acromegaly: results of the ACROSPECT study. Eur J Endocrinol 2014; 170: 791-797.

(10) Kasuki L, Marques NV, Nuez MJ, Leal VL, Chinen RN, Gadelha MR. Acromegalic patients lost to follow-up: a pilot study. Pituitary 2013;16:245-250.

(11) Andemariam B, Owarish-Gross J, Grady J, Boruchov D, Thrall RS, Hagstrom JN. Identification of risk factors for an unsuccessful transition from pediatric to adult sickle cell disease care. Pediatr Blood Cancer 2014;61(4):697-701.

(12) Wojciechowski EA, Hurtig A, Dorn L. A natural history of adolescents and young adults with sickle cell disease as they transfer to adult care: a need for case management services. J Pediatr Nurs 2002;17(1):18-27.

(13) Hazel E, Zhang X, Duffy CM, Campillo S. High rates of unsuccessful transfer to adult care among young adults with juvenile idiopathic arthritis. Pediatr Rheumatol Online J 2010; 8(2):1-6.

(14) Attanasio R, Montini M, Valota M, Cortesi L, Barbo R, Biroli F et al. An audit of treatment outcome in acromegalic patienst attending our center at Bergamo, Italy. Pituitary 2008; 11:1-11.
(15) Lanoy E, Mary-Krause M, Tattevin P, Dray-Spira R, Duvivier C, Fisher P et al. Predictors identified for losses to follow-up among HIV-seropositive patients. J Clin Epidemiol 2006 ;59:829-835.
(16) Bowen ME, Henske JA, Potter A. Health Care Transition in Adolescents and Young Adults with Diabetes. Clinical Diabetes 2010;28(3):99-106.
(17) Yeung E, Kay J, Roosevelt GE, Brandon M, Yetman AT. Lapse of care as a predictor for morbidity in adults with congenital heart disease. Int J Cardiol 2008;125(1):62-65.
(18) Goossens E, Stephani I, Hilderson D, Gewillig M, Budts W, Van Deyk K et al. Transfer of Adolescents With Congenital Heart Disease From Pediatric Cardiology to Adult Health Care. An Analysis of Transfer Destinations. J Am Coll Cardiol 2011;57(23):2368-2374.
(19) Reid GJ, Irvine MJ, McCrindle BW, Sananes R, Ritvo PG, Siu SC et al. Prevalence and correlates of successful transfer from pediatric to adult health care among a cohort of young adults with complex congenital heart defects. Pediatrics 2004;113(3Pt1):e197-e205.
(20) Mackie AS, Rempel GR, Rankin KN, Nicolas D, Magill-Evans J. Risk factors for loss to follow-up among children and young adults with congenital heart disease. Cardiol Young 2012;22(3):307-315.
(21) Wacker A, Kaemmerer H, Hollweck R, Hauser M, Deutsch MA, Brodherr-Heberlein S et al. Outcome of operated and unoperated adults with congenital cardiac disease lost to follow-up for more than five years. Am J Cardiol 2005;95(6):776-779.
(22) de Bono J, Freeman LJ. Aortic coarctation repair - lost and found: The role of local long term specialised care. Int J Cardiol 2005;104(2):176-183.
(23) Gleeson H, Turner G. Transition to adult services. Arch Dis Child Educ Pract Ed 2012;97(3) :86-92.
(24) Betz CL. Transition of adolescents with special healthcare needs: review and analysis of the literature. Issues Compr Pediatr Nurs 2004;27(3):179-241.
(25) Crowley R, Wolfe I, Lock K, McKee M. Improving the transition between paediatric and adult healthcare: a systematic review. Arch Dis Child 2011;(96):548-553.
(26) Jalkut MK, Allen PJ. Transition from pediatric to adult health care for adolescents with congenital heart disease: a review of the literature and clinical implications. Ped Nurs 2009; 35(6):381-387.
(27) Stewart D, Stavness C, King G et al. A critical appraisal of literature reviews about the transition to adulthood for youth with disabilities. Phys Occup Ther 2006;26:5-24.
(28) Goossens E, Fleck D, Canobbio MM, Harrison JL, Moons P. Development of an international research agenda for adult congenital heart disease nursing. Eur J Cardiovasc Nurs 2013; 12(1):7-16.
(29) Oster ME, Riehle-Colarusso T, Simeone RM, Gurvitz M, Kaltman JR, McConnell M et al. Public health science agenda for congenital heart defects: report from a Centers for Disease Control and Prevention Experts Meeting. J Am Heart Assoc 2013; 2(5):e000256.
(30) Cotts T, Khairy P, Opotowsky AR, John AS, Valente AM, Zaidi AN et al. Clinical research priorities in adult congenital heart disease. Int J Cardiol 2014;171(351):360.
(31) Mackie AS, Ionescu-Ittu R, Therrien J, Pilote L, Abrahamowicz M, Marelli AJ. Children and adults with congenital heart disease lost to follow-up: who and when? Circulation 2009;120:302-309.
(32) Iversen K, Vejlstrup NG, Sondergaard L, Nielsen OW. Screening of adults with congenital cardiac disease lost to follow-up. Cardiol Young 2007;17:601-608.
(33) Wray J, Frigiola A, Bull C, Adult Congenital Heart disease Research Network (ACoRN). Loss to specialist follow-up in congenital heart disease; out if sight, out of mind. Heart 2013; 99(7):485-490.

(34) Norris MD, Webb GD, Drotar D, Lisec A, Pratt J, King E et al. Prevalence and patterns of retention in cardiac care in young adults with congenital heart disease. J Pediatr 2013; 163(3):902-904.
(35) Reiss JG, Gibson RW, Walker LR. Health care transition: youth, family, and provider perspectives. Pediatrics 2005;115(1):112-120.
(36) Mylotte D, Pilote L, Ionescu-Ittu R, Abrahamowicz M, Khairy P, Therrien J et al. Specialized Adult Congenital Heart Disease Care: The Impact of Policy on Mortality. Circulation 2014;129(18):1804-1812.
(37) Norris MD, Webb GD, Drotar D, Lisec A, Pratt J, Akanbi F et al. Risk factors associated with loss to follow-up in young adults with congenital heart disease. J Am Coll Cardiol 2012;59(13s1):E790.
(38) US Census Bureau 2ACS. State-to-State Migration Flows: 2012 American Community Survey 1-Year Estimates. 2012. US Census Bureau.
(39) Algemene Directie Statistiek en Economische Informatie. Kerncijfers 2012. Statistisch overzicht van België. S000.01N/2012, 1-58. 2012. Brussels, Belgium, FOD Economie, KMO, Middenstand en Energie.
(40) Wren C, O'Sullivan JJ. Loss to follow-up of adults with repaired congenital heart disease. Heart 2013;99(7):440-441.
(41) Baumgartner H, Bonhoeffer P, De Groot NMS, de Haan F, Deanfield J, Galie N et al. ESC Guidelines for the management of grown-up congenital heart disease (new version 2010). Eur Heart J 2001;31(23):2915-2957.
(42) Landzberg MJ, Murphy DJ, Davidson WR, Jarcho JA, Krumholz HM, Mayer JE et al. Task Force 4: Organization of Delivery Systems for Adults with Congenital Heart Disease. J Am Coll Cardiol 2001;37(5):1187-1193.
(43) Paulus D, Van Den Heede K, Mertens R. Organisatie van zorg voor chronisch zieken in België: ontwikkeling van een position paper. KCE Reports 190A. D/2012/10.273/79., 1-184. 2012. Health Services Research (HSR). Brussel: Federaal Kenniscentrum voor de Gezondheidszorg (KCE).
(44) Enthoven AC. Integrated Delivery Systems: The Cure for Fragmentation. Am J Manag Care 2009;15:S284-S290.
(45) Wagner E, Austin BT, Van Korff M. Organizing Care for Patients with Chronic Illness. Milbank Q 1996;74:511-514.
(46) Starfield B. Is primary care essential? Lancet 1994;344:1129-1133.
(47) Starfield B. Primary care: concept, evaluation and policy. New York: Oxford university Press 1992.
(48) van Staa A, van der Stege HA, Jedeloo S, Moll HA, Hilberink SR. Readiness to Transfer to Adult Care of Adolescents with Chronic Conditions: Exploration of Associated Factors. J Adolesc Health 2011;48(3):295-302.
(49) Cetta F, Warnes CA. Adults with congenital heart disease: patient knowledge of endocarditis prophylaxis. Mayo Clin Proc 1995;70(1):50-54.
(50) Kantoch MJ, Collins-Nakai RL, Medwid S, Ungstad E, Taylor DA. Adult patients' knowledge about their congenital heart disease. Can J Cardiol 1997;13(7):641-645.
(51) Knirsch W, Hassberg D, Beyer A, Teufel T, Pees C, Uhlemann F et al. Knowledge, compliance and practice of antibiotic endocarditis prophylaxis of patients with congenital heart disease. Pediatr Cardiol 2003;24(4):344-349.
(52) Moons P, De Volder E, Budts W, De Geest S, Elen J, Waeytens K et al. What do adult patients with congenital heart disease know about their heart disease, treatment, and prevention of complications? Heart 2001;86(1):74-80.

(53) Van Deyk K, Pelgrims E, Troost E, Goossens E, Budts W, Gewillig M et al. Adolescents' understanding of their congenital heart disease on transfer to adult-focused care. Am J Cardiol 2010;106(12):1803-1807.
(54) Goossens E, Van Deyk K, Zupancic N, Budts W, Moons P. Effectiveness of structured patient education on the knowledge level of adolescents and adults with congenital heart disease. Eur J Cardiovasc Nurs 2014;13(1):63-70.
(55) Van Deyk K, Moons P, Farroni N, Dedroog D, Pinxten S, Troost E. Effects of a structured education programme for adult patients with congenital heart disease: a follow-up study. Acta Cardiologica 2008:63(1):125-126.
(56) Rönning H, Nielsen NK, Strömberg A, Thilen U, Swahn E. Development and evaluation of a computer-based educational program for adults with congenitally malformed hearts. Eur J Cardiovasc Nurs 2012;12(1):78-86.
(57) Brook JS, Zhang C, Brook DW. Developmental trajectories of marijuana use from adolescence to adulthood: personal predictors. Arch Pediatr Adolesc Med 2011;165(1):55-60.
(58) From the Centers for Disease Control and Prevention. Health-risk behaviors among persons aged 12-21 years-United States, 1992. JAMA 1994;271(17):1311.
(59) Kolbe LJ. An epidemiological surveillance system to monitor the prevalence of youth behaviors that most affect health. Health Education 1990;21:44-48.
(60) Suris JC, Michaud PA, Akre C, Sawyer SM. Health risk behaviors in adolescents with chronic conditions. Pediatrics 2008;122(5):e1113-e1118.

11 LAY SUMMARY – SAMENVATTING

Lay summary

Congenital heart disease (CHD) is the most common birth defect in newborns, affecting about 9 per 1,000 babies worldwide. It comprises a wide spectrum of anatomical defects of the heart and/or great intrathoracic vessels that have a variable long-term impact on patients' health status, functionalities, and overall well-being. While CHD was one of the most common causes of infant mortality around the 1960s, about 90% of children with CHD nowadays have the prospect of reaching adult age. Over the past decades, tremendous investments have been made in the care for patients with CHD through the optimization of CHD detection, surgery, and long-term management. However, despite this ameliorated life-expectancy, patients with CHD have a life-time risk for developing life-threatening complications such as heart failure, arrhythmias, pulmonary hypertension, endocarditis, and sudden cardiac death. In order to prevent such serious complications, the provision of life-long follow-up care performed by specialized healthcare professionals at predefined time intervals is recommended by experts.

Since the life expectancy of patients with CHD increased substantially, CHD became a life cycle disease. Patients have the prospect to reach several phases of life such as adolescence, adulthood, and parenthood. However, in order to address the specific healthcare needs of patients at each of these life stages, age-appropriate care programs are required. During childhood, patients should receive care at specialized pediatric cardiology programs. When reaching adulthood, patients are advised to transfer their follow-up care to adult-focused facilities. Based on the type of heart defects, patients will be recommended to transfer to either adult congenital heart disease (ACHD) programs, non-specialist general cardiology care settings, or shared care programs. Such transfers of care across different healthcare settings require patients to re-adapt themselves to a new context, healthcare team, responsibilities, and requirements. For some patients such handing-off of care could be stressful and be associated with detrimental outcomes. Understandably, these complex transfers of care across settings make patients vulnerable for a discontinuation of their care process. Such discontinuations characterized by exceeding the recommended guideline-based time interval between recommended follow-up visits are called 'care gaps'.

In order to prepare, guide, and support patients for these transfers of care, comprehensive care programs facilitating this transition towards a new life phase, healthcare context, and set of responsibilities is recommended by international experts. Such care programs provide comprehensive care to patients addressing both medical and psychosocial needs of patients.

As part of this PhD project on transitional care in young people with CHD, a total of eight studies were performed covering two important and highly relevant themes: the importance of life-long care in patients with CHD, and the provision of transitional care in young people with CHD.

Although it is very important for patient to have regular check-ups of their cardiovascular and overall health status, this PhD demonstrated that care gaps are highly prominent in young people with CHD transferring to adult care. However, when comparing results of different studies from an international perspective, there appeared to be a significant difference in the magnitude of this problem across healthcare systems. Furthermore, important differences were found between the number of young people remaining in pediatric cardiology, discontinuing cardiac follow-up care, or becoming completely untraceable within the healthcare system. Based on a review of the existing scientific literature regarding the problem of care gaps in the overall population of young people diagnosed with a chronic condition, several risk and protective factors were identified. Four categories of determinants were identified: demographics, disease-related characteristics, healthcare services use, and patient's health behavior. Additional studies are, however, needed to identify other determinants and ultimately develop effective strategies that can prevent patients from experiencing care gaps during life.

As young people with CHD develop towards becoming adults, they are supposed to attain a set of skills, insights, attitudes, and health behaviors required in adult life and healthcare. The provision of transitional care as a comprehensive package of interdisciplinary guidance, support, and care is recommended by experts. Patient education on a broad range of CHD-related aspects is recommended as an important component of this transitional care. This PhD clearly demonstrated significant gaps in the knowledge of patient regarding their condition, treatment, preventive measures, and implications of the condition in adult life. This PhD demonstrated that the provision of one structured, tailored education session results in a significant but small improvement of the disease-related knowledge. No improvement of health behaviors was, however, demonstrated in this study. Innovative supplementary interventions aiming to increase patients' level of disease-related knowledge must be developed and tested in the future.

Adolescents typically engage in a more risky lifestyle as part of adolescent experimental behaviors with the use of tobacco, alcohol, and illicit drugs. Since a heart-healthy lifestyle is very important for patients with CHD, educational interventions informing patients about these risks and explain which lifestyle is deemed mandatory. The prevalence of health-compromising behaviors appeared to be fairly low in our sample of more than 400 adolescents with CHD. These data, however, demonstrated a significant increase of experimenting behaviors around the age at which

patients generally transfer to ACHD care (i.e., 16 years). This finding indicated a need for actively detecting risky behaviors in patients. This can be done in a valid way by using the HBS-CHD. Altering the health behaviors of young people through the provision of education seemed, however, challenging and rather ineffective. This PhD, however, could not demonstrate a beneficial effect of education on the correction of health risk behaviors of young people with CHD. Future studies are needed in order to investigate if there is a potential effect when patients are exposed to multiple subsequent educational sessions over a longer time span.

Samenvatting

Een aangeboren hartaandoening (AHA) is de meest voorkomende aangeboren afwijking vastgesteld bij pasgeborenen. Deze aandoening treft wereldwijd ongeveer 9 per 1000 pasgeborenen. AHA omvat een brede verzameling van anatomische afwijkingen van het hart en/of de grote intrathoracale bloedvaten dewelke een variabele impact hebben op de gezondheidsstatus, de functioneringsgraad en het algemeen welzijn van patiënten. Ondanks de vaststelling dat AHA de meest voorkomende oorzaak van kindersterfte was in de jaren ’60, bereikt heden ongeveer 90% van de kinderen geboren met AHA de volwassen leeftijd. Doorheen de laatste decaden werden er significante investeringen gedaan in de zorg voor patiënten met AHA. Deze inspanningen leidde tot een verbeterde detectie van AHA, betere chirurgische behandelingstechnieken en een verbeterde lange termijnsvisie op het ziektemanagement. Ondanks deze verbeterde vooruitzichten in termen van levensverwachting, hebben patiënten met AHA echter een levenslang risico op de ontwikelling van levensbedreigende complicaties zoals hartfalen, hartritmestoornissen, pulmonale hypertensie, endocarditis en plotse cardiale dood. Om dergelijke complicaties te voorkomen wordt een levenslange follow-up, uitgevoerd door gespecialiseerde gezondheidswerkers op regelmatige tijdstippen, aanbevolen door experten.

Doordat de levensverwachting van patiënten met AHA sterk verbeterde is deze aandoening een chronische aandoening geworden die doorheen het ganse leven van de patiënt aanwezig blijft. Patiënten hebben heden het vooruitzicht om diverse levensfasen te bereiken zoals adolescentie, volwassenheid en ouderschap. Echter om de specifieke zorgnoden van deze patiënten met AHA doorheen de verschillende opeenvolgende levensfasen te kunnen beantwoorden, moeten er leeftijdsgebonden zorgprogramma’s ontwikkeld worden. Tijdens de kindertijd worden kinderen met AHA opgevolgd op gespecialiseerde afdelingen voor kindercardiologie. Echter wanneer zij de volwassen leeftijd bereiken, worden adolescenten geadviseerd om hun cardiale follow-up te transfereren naar zorgprogramma’s voor volwassenen met AHA. Op basis van de gediagnosticeerde aangeboren hartafwijking krijgen patiënten het advies om hun zorg te transfereren naar een gespecialiseerd zorgprogramma voor volwassenen met AHA, een niet-gespecialiseerde algemene cardioloog, of naar een programma waar zorg gedeeltelijk wordt uitgevoerd door gespecialiseerde en gedeeltelijk door niet-gespecialiseerde cardiologen (shared care). Dergelijke transfers van zorg over verschillende settings heen impliceert dat patiënten zich steeds moeten aanpassen aan een nieuwe context, een nieuwe zorgteam, en gerelateerde verwachtingen. Voor sommige patiënten is deze transfer stresserend en resulteert het in nadelige uitkomsten. Dergelijke transfers van zorg maken patiënten namelijk kwetsbaar voor discontinuering van het zorgproces. Een dergelijke onderbreking

van het zorgprocess gekenmerkt door het overschrijden van de aanbevolen termijn tussen follow-up controles wordt ook wel omschreven als ‘care gaps’.

Om patiënten voor te bereiden, te begeleiden en te ondersteunen tijdens dergelijke transfers van zorg is het aanbevolen om een allomvattend zorgprogramma aan te bieden. Een dergelijk transitieprogramma heeft tot doel om de overgang naar een nieuwe levensfase, gezondheidszorg-context en set van verantwoordelijkheden te faciliteren. Dergelijke transitieprogramma’s bieden een breed zorgpakket aan patiënten waarbij zowel medische als psychosociale zorgnoden worden beantwoord.

Als onderdeel van dit doctoraatsonderzoek naar transitionele zorg voor jongeren met AHA werden er in totaal acht studies uitgevoerd naar twee belangrijke en klinisch relevante onderwerpen: het belang van levenslange opvolging van patiënten met AHA en het aanbieden van transitionele zorg aan jongeren met AHA.

Ondanks het feit dat het voor patiënten met AHA enorm belangrijk is om op regelmatige basis een controle te laten uitvoeren van de cardiovasculaire en algemene gezondheid, toonde dit doctoraat aan dat onderbrekingen van het zorgproces (care gaps) prominent optreden bij jongeren met AHA tijdens de transfer naar zorgprogramma’s voor volwassenen met AHA. Echter wanneer we de resultaten van verschillende studies omtrent deze problematiek met elkaar vergelijken, zien we grote verschillen in de grootte van dit probleem tussen verschillende gezondheidszorgsystemen. Daarenboven werden er ook belangrijke verschillen geobserveerd in het aantal jong-volwassenen die op de kindercardiologische afdeling blijven, cardiale follow-up onderbreken, of zelfs ondetecteerbaar werden in het gehele gezondheidszorgsysteem. Op basis van een literatuurstudie betreffende het probleem van zorgonderbrekingen in de algemene populatie van jongeren met een chronische aandoening werden er zowel risico- als protectieve factoren geïdentificeerd. Vier categorieën van determinanten werden geïdentificeerd: demografische kenmerken, kenmerken gerelateerd aan de aandoening, gezondheidszorggebruik, en het gezondheidsgedrag van patiënten. Bijkomende studies zijn echter noodzakelijk om bijkomstige determinanten van discontinuering van zorg te identificeren en uiteindelijk ook effectieve strategieën te ontwikkelen die een dergelijke discontinuering kan voorkomen.

Wanneer jongeren met AHA ontwikkelen naar opgroeiende volwassenen, worden zij verondersteld om een set van vaardigheden, inzichten en attitudes eigen te maken die vereist zijn in het volwassen leven en zorgsysteem. Het aanbieden van transitionele zorg als een allomvattend pakket van interdisciplinaire ondersteuning, begeleiding en zorg wordt sterk aanbevolen door

internationale experten. Patiënteneducatie betreffende diverse AHA-gerelateerde aspecten wordt aanbevolen als een belangrijke component van transitionele zorg. Dit doctoraatsonderzoek toonde aan dat er significante tekorten zijn in de kennis van patiënten omtrent hun aandoening, behandeling, preventieve gedragingen en implicaties van de aandoening op het verdere volwassen leven. Dit doctoraat toonde aan dat één gestructureerde, geïndividualiseerde educatiesessie resulteert in een significante, doch kleine verbetering van het kennisniveau. Een significante verbetering van het gezondheidsgedrag kon echter niet worden aangetoond in dit doctoraat. Innovatieve supplementaire interventies die tot doel hebben om het niveau van kennis te verhogen bij patiënten met AHA moeten in de toekomst verder ontwikkeld en getest worden.

Adolescenten vertonen typisch risicovoller gezondheidsgedrag als onderdeel van hun ontwikkeling als adolescent. Adolescentie is namelijk gekenmerkt door experimenteel gebruik van tabak, alcohol en (illegale) drugs. Het aanmeten van een hartbeschermende levensstijl is echter van enorm groot belang voor patiënten met AHA. Het geven van educatie aan patiënten over de risico's van een risicovolle levensstijl en het omschrijven van het gewenste preventieve gedrag wordt belangrijk geacht door experten en clinici. Dit doctoraat toonde een significante toename van risicovol gezondheidsgedrag aan bij adolescenten net op de leeftijd waar zij doorgaans de transfer naar volwassen zorg maken, namelijk rond de leeftijd van 16j. Deze bevinding duidt aan dat het zinvol is om op een actieve wijze risicovol gedrag te detecteren bij patiënten met AHA. Een dergelijke detectie kan op valide wijze worden uitgevoerd aan de hand van de 'Gezondheidsgedragschaal voor Aangeboren Hartaandoeningen'. Het wijzigen van risicovol gezondheidsgedrag bij jongeren door middel van educatie lijkt echter een uitdagende en eerder ineffectieve strategie te zijn. Dit doctoraat kon namelijk geen positief effect aantonen in het verbeteren van gezondheidsgedrag bij jongeren met AHA door educatie. Toekomstige studies zijn noodzakelijk om te onderzoeken of een groter effect kan bereikt worden indien patiënten worden blootgesteld aan meerdere opeenvolgende educatiesessies over een langere tijdsperiode

12 CURRICULUM VITAE

Eva Goossens was born on May 15, 1985, in Bonheiden, Belgium. She graduated as a Bachelor in Nursing at the Catholic College University Leuven in 2007 (Summa cum Laude) and obtained a Master of Nursing and Obstetrics at KU Leuven in 2009 (Magna cum Laude). She holds a certificate in Case Management in Acute Hospitals of the Center for Health Services and Nursing Research (KU Leuven).

In 2009, she started working as a Research Associate at the Center for Health Services and Nursing Research (KU Leuven). She worked as a co-investigator and project coordinator on several research projects related to geriatric and psychiatric care performed by order of the Ministry of Public Health. In January 2010, Eva started her doctoral training in Biomedical Science at the doctoral school of Patient-related and Public Health Research at the Faculty of Medicine, KU Leuven on transitional care for young people with congenital heart disease. In October 2011, she obtained a PhD fellowship at the Research Foundation Flanders (FWO) as the very first Master in Nursing and Obstetrics at the KU Leuven. As part of her PhD project, she coordinated several studies in collaboration with researchers from Boston (USA), Copenhagen (Denmark), and Bergen (Norway), and led an international study under the auspices of the International Society of Adult Congenital Heart Disease (ISACHD) Nursing Network. Furthermore, Eva was a visiting PhD fellow at the McGill Adult unit for Congenital Heart Disease Excellence (MAUDE), McGill University, Montréal, Canada.

During her PhD training, Eva received the 'Best Poster Prize' at the 7th International IV Therapy conference for her master's thesis on sensory perceptions of patients during surgical insertion of a totally implantable venous access device. Furthermore, she received an award for the 'Best Moderated Poster Presentation' on the 12th Annual Spring Meeting on Cardiovascular Nursing. In 2014, she was elected by the Editorial Board of the European Journal of Cardiovascular Nursing to receive the 'Top 10 reviewer award'. Eva became member of the Council on Cardiovascular Nursing and Allied Professionals (CCNAP) in 2011 and the Belgian Working Group on Cardiovascular Nursing (BWGCVN) in 2013. In 2014, she was elected as Vice-Chair of the BWGCVN.

Finally, she has appointments as a guest lecturer in Pediatric Nursing at Thomas More, UCLeuven-Limburg, and VIVES.

25 Moceri P, **Goossens E,** Hascoet S, Checler C, Bonello B, Ferrari E, Acar P, Fraisse A. (2015) From adolescence to adults with congenital heart disease: the role of transition. European Journal of Pediatrics (Epub ahead of print).

24 **Goossens E**, Fernandes SM, Landzberg MJ, Moons P. (2015) Implementation of the American College of Cardiology/American Heart Association 2008 Guidelines for the Management of Adults with Congenital Heart Disease. American Journal of Cardiology (Epub ahead of print, doi : 10.1016/j.amjcard.2015.04.041).

23 **Goossens E**, Fieuws S, Van Deyk K, Luyckx K, Gewillig M, Budts W, Moons P. (2015). Effectiveness of structured education on knowledge and health behaviors in patients with congenital heart disease. The Journal of Pediatrics (Epub ahead of print, doi : 10.1016/j.jpeds.2015.02.041).

22 Apers S, Luyckx K, **Goossens E**, Rassart J, Budts W, Moons P. (2015). Sense of coherence in young people with congenital heart disease. Journal of Developmental & Behavioral Pediatrics, 36(4):267-76.

21 Janssens A, **Goossens E**, Luyckx K, Budts W, Gewillig M, Moons P, for the i-DETACH investigators (2014). Exploring the relationship between disease-related knowledge and health risk behaviours in young people with congenital heart disease. European Journal of Cardiovascular Nursing (Epub ahead of print).

20 Luyckx K, **Goossens E**, Rassart J, Apers S, Vanhalst J, Moons P. (2014) Parental support, internalizing symptoms, perceived health status, and quality of life in adolescents with congenital heart disease: influences and reciprocal effects. Journal of Behavioral Medicine, 37(1): 145-55.

19 **Goossens E**, Van Deyk K, Zupancic N, Budts W, Moons P. (2014) Effectiveness of structured patient education on the knowledge level of adolescents and adults with congenital heart disease. European Journal of Cardiovascular Nursing, 13(1): 63-70.

18 Rassart J, Luyckx K, **Goossens E**, Apers S, Moons P. (2014) A closer look at the developmental interplay between parenting and perceived health in adolescents with congenital heart disease. Journal of Behavioral Medicine, 37(6): 1202-14.

17 Vanhalst J, Rassart J, Luyckx K, **Goossens E**, Apers S, Goossens L, Moons P. (2013) Trajectories of Loneliness in Adolescents with Congenital Heart Disease: Associations With Depressive Symptoms and Perceived Health. Journal of Adolescent Health, 53(3):342-49.

16 Apers S, Luyckx K, Rassart J, **Goossens E**, Budts W, Moons P. (2013) Sense of coherence is a predictor of perceived health in adolescents with congenital heart disease: A cross-lagged prospective study. International Journal of Nursing Studies, 50(6):776-78.

15 Apers S, Moons P, **Goossens E**, Luyckx K, Gewillig M, Bogaerts K, Budts W. (2013) Sense of coherence and perceived physical health explain the better quality of life in adolescents with congenital heart disease. European Journal of Cardiovascular Nursing, 12(5):475-83.

14 Hilderson D, Westhovens R, Wouters C, Van der Elst K, **Goossens E**, Moons, P. (2013) Rationale, design and baseline data of a mixed methods study examining the clinical impact of a brief transition programme for young people with juvenile idiopathic arthritis: the DON'T RETARD project. BMJ Open, 3 (12): e003591.

13 Rassart J, Luyckx K, **Goossens E**, Apers S, Klimstra T, Moons P. (2013) Personality traits, quality of life, and perceived health in adolescents with congenital heart disease. Psychology & Health, 28(3):319-35.

12 **Goossens E**, Luyckx K, Mommen N, Gewillig M, Budts W, Zupancic N, Moons P. (2013) Health risk behaviors in adolescents and emerging adults with congenital heart disease: Psychometric properties of the Health Behavior Scale-CHD. European Journal of Cardiovascular Nursing, 12:544-57.

11 **Goossens E**, Fleck D, Canobbio M, Harrison J, Moons P, on behalf of the International Society of Adult Congenital Heart Disease (ISACHD) Nursing Network (2013) Development of an international research agenda for adult congenital heart disease nursing. European Journal of Cardiovascular Nursing, 12(1): 7-16.

10 Luyckx K, **Goossens E**, Apers S, Rassart J, Klimstra T, Dezutter J, Moons P. (2012) The 13-item sense of coherence scale in Dutch-speaking adolescents and young adults: Structural validity, age trends, and chronic disease. Psychologica Belgica, 52(4): 351-68.

9 Luyckx K, Missotten L, **Goossens E**, Moons P. (2012) Individual and contextual determinants of quality of life in adolescents with congenital heart disease. Journal of Adolescent Health, 51(2):122-8.

8 Rassart J, Luyckx K, Apers S, **Goossens E**, Moons P. (2012) Identity dynamics and peer relationship quality in adolescents with a chronic disease: the sample case of congenital heart disease. Journal of Developmental & Behavioral Pediatrics, 33(8): 625-32.

7 **Goossens E**, Hilderson D, Moons P. (2012) Coaching through transition: A challenge for critical care nurses. Australian Critical Care, 25(1): 1-2.

6 **Goossens E**, Stephani I, Hilderson D, Gewillig M, Budts W, Van Deyk K, Moons P. (2011) Transfer of adolescents with congenital heart disease from pediatric cardiology to adult health care: an analysis of transfer destinations. Journal of the American College of Cardiology, 57(23): 2368-74.

5 **Goossens E**, Norekval T, Faerch J, Hody L, Olsen S, Darmer M, Jaarsma T, Moons P. (2011) Sexual counseling of cardiovascular patients in Europe: Culture matters. International Journal of Clinical Practice, 65(10): 1092-99.

4 **Goossens E**, Goossens G, Stas M, Janssens C, Jérôme M, Moons P. (2011) Sensory perceptions of oncology patients undergoing surgical insertion of a totally implantable venous access device: a qualitative exploratory study. Oncology Nursing Forum, 38(1): E20-E26.

3 Luyckx K, **Goossens E**, Van Damme C, Moons P. (2011) Identity formation in adolescents with congenital cardiac disease: A forgotten issue in the transition to adulthood. Cardiology in the Young, 21(4): 411-20.

2 Luyckx K, **Goossens E**, Missotten L, Moons P, the i-DETACH investigators (2011) Adolescents with congenital heart disease: The importance of perceived parenting for psychosocial and health outcomes. Journal of Developmental & Behavioral Pediatrics, 32(9): 651-9.

1 Van Deyk K, Pelgrims E, Troost E, **Goossens E**, Budts W, Gewillig M, Moons P. (2010) Adolescents' understanding of their congenital heart disease upon the transfer to adult-focused care. American Journal of Cardiology, 106(12): 1803-7.

Articles in other scientific journals

4 **Goossens E**, Apers S, Moons P. (2014) Qualité de vie des patients atteints de cardiopathies univentriculaires. Le Livret du XXXVème Séminaire de Cardiologie Congénitale et Pédiatrique.

3 Apers S, **Goossens E**, Dancet E, Luyckx K, Moons P. (2013) Psychologie en verloskunde: het belang van 'sense of coherence'. Tijdschrift voor Vroedvrouwen, 19(5): 310-3.

2 **Goossens E**, Moons P. (2013) Levenslange zorg voor patiënten met een aangeboren hartafwijking. Tijdschrift voor Cardiologie, 25(1): 7-13.

1 **Goossens E**, Moons P. (2013) Une vie de soins pour les patients souffrant d'une malformation cardiaque congénitale. Journal de Cardiologie, 25(1): 7-13.

Book chapters in professionally oriented books published by international publishers

2 **Goossens E**, Kovacs A, Mackie A, Moons P. (2013) Transfer and transition in congenital heart disease. In: da Cruz E, Ivy D, Jaggers J, Hraska V. (Eds.)*, Pediatric Cardiology, Cardiac Surgery and Intensive Care*, Chapt. 11. London, United Kingdom: Springer-Verlag.

1 **Goossens E**, Moons P, Meijboom FJ. (2013) Transitie van jongvolwassenen. [Transitional care for young adults] In: Mulder BJM, Pieper PG, Meijboom FJ, Hamer JPM (Eds.), Aangeboren hartafwijkingen bij volwassenen [Congenital Heart Disease in Adults], Chapt. 3. Houten, The Netherlands: Bohn Stafleu van Loghum Springer media.